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A TREATISE

ON THE

NERVOUS DISEASES OF CHILDREN

FOR

PHYSICIANS AND STUDENTS

BY

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PREFACE TO SECOND EDITION.

THE author has reason to be pleased with the praise bestowed upon the first edition of his book in this country and in Europe. Translations into German and Italian have helped to extend its teachings, and a French edition is to appear shortly. Recent writers of text-books on Medicine and on Pediatrics have made liberal use of it. It is evident that the book has filled a distinct gap in medical literature.

In preparing the second edition the writer has endeavored to reduce the size of the book without lessening its value as a special treatise on the nervous diseases of children. He has omitted the chapters on Anatomy and Physiology, the subject matter of which can readily be found in the text-books of Dana, Gowers, Starr and Oppenheim. All detailed histories of cases and all bibliographies have been omitted. The names of authors have, however, been inserted in the text to give credit for work done,—a bit of justice which a writer should be happy and ready to do to others.

Every chapter has been carefully revised and much new matter has been introduced. A comparison of the two editions of this book will show that important advances have been made in Neurology and the allied sciences within recent years.

The author wishes to express his especial thanks to his publishers for their interest in this new edition, and to Dr. Alfred Wiener, of this City, for his assistance in preparing the index.

21 EAST SIXTY-FIFTH STREET.

June, 1905.

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THE NERVOUS DISEASES OF CHILDREN.

CHAPTER I.

INTRODUCTION—METHODS OF EXAMINATION.

THE nervous system of the child is subject to many diseases. Some of these are identical with the nervous disorders of tne adult; others are peculiar to the early years of life.

The brain and the spinal cord do not attain their full development until months and years after birth, and even the peripheral nerves do not exhibit all their normal functions until the child is several weeks old. During this period of incomplete development, the nervous system responds much more readily to morbid influences than it does in later years. This is especially true of the brain. It is in a state of irritability and instability, and a perversion of functions may result from causes which would exert little or no influence over the nervous system of the youth or adult. Evidence of this is furnished by the behavior of a child in fever. The irregular choreiform twitchings and the delirium are often the outward signs of an unstable cerebral state, while the unusual irritability of the brain is proved by the occurrence of convulsions upon peripheral irritation to which the adult brain would not at all respond.

In the early period of life, too, hereditary affections of the nervous system are frequently manifest, and morbid psychic inheritance casts its shadows before. Inhibition of normal development may occur at any period; family affections are developed in the earlier years of life, and the acute infectious diseases of childhood are often followed by serious nervous disorders. If we add to these, diseases due to traumatism, we have an array of nervous disorders equal

to, if not in excess of, those that occur after the age of puberty. Childhood is exempt only from the diseases due to senile deterioration, and is relatively free from those due to toxic agents, such as alcohol, metallic poisons, and syphilis; but the effect of such diseases in the parent is exhibited with cruel persistence in the offspring.

The diseases of the nervous system during the period of incomplete development are to be the special subject of this treatise. Before proceeding to the description of disease it will be necessary to adopt a correct

METHOD OF EXAMINATION.—First of all inquire into the ancestry of the child. A reliable history of the physical and mental condition of parents, grandparents, and other relatives is of the greatest value in establishing a diagnosis. The habits and the diseases of the parents should be carefully determined, for of the ills the child is heir to, not a few are due to alcoholism, to syphilis, to epilepsy, to hysteria, to psychic disorders of a parent. Next to heredity, environment plays the most important part; it is well, therefore, to inquire into the home surroundings, the manner in which the child is watched and cared for; how it has been fed, trained, and educated.

The previous history of the child is next in order. In every case inquire into the manner of its birth; whether it was protracted or not; whether or not instruments were used; whether the child was asphyxiated at birth or at once began to breathe freely. Make inquiries regarding the occurrence of spasms or convulsions; the time at which the child began to take notice of things, to recognize parent or nurse, to stand, to walk, and to talk. The occurrence of other diseases, of the ordinary infectious diseases of childhood, of whooping-cough, of pneumonia, of scarlet fever, of measles and meningitis, should be determined, and one should never forget to ask whether similar nervous conditions have been previously observed. Then proceed to the

EXAMINATION OF THE PATIENT.—The art of making a diagnosis by mere inspection has gone out of date, and is cried down by many; yet I am willing to say that in fully one-half of the nervous diseases of children the nature of

the trouble can be suspected, if not made out, by a thorough inspection of the child without putting a finger to its body. I am not in favor of hurried examinations; on the contrary, I wish to plead for the greatest accuracy in examining for details; but let the physician or student train his powers of observation and his diagnostic ability will be more acutely developed than that of the man who can never even suspect a disease unless he has all his tools (percussion-hammer, thermometer, æsthesiometer, electrodes) constantly at his command.

In my lectures to students I insist that they shall study the general appearance of a child, and should not feel satisfied until they learn to recognize peculiarities of facial expression, of gait, and of stature; to distinguish between the behavior of the normal child and the feeble-minded, between spastic and flaccid palsies, and to determine by the peculiar deformity of the foot or by the scraping noise which the patient makes in walking, which group of muscles is affected. It is important from the history of the patient, and from these general observations to get correct first impressions; these first impressions are then to be confirmed by a careful detailed examination.

Never make a diagnosis unless the child has been wholly undressed; if this is not done a Pott's paralysis may be taken to be a traumatic myelitis, or a neuritis may be mistaken for poliomyelitis anterior. Lay the child on a table or on another person's lap in order to get a full view of it; of the relative size of head and body; of the proportionate development of arms, legs, and abdomen. Remember also that the child has heart and lungs, liver, spleen, and intestines, which, if diseased, may hold an important relation to the nervous disorder present. In proceeding to a detailed examination it is best to begin with the head, including the face, then take up the upper extremities, the abdomen, and finally the lower extremities.

The following scheme includes the more important points to be established in the examination of a child; the exact order of inquiry is subject to slight modifications.

EXAMINATION SCHEME.

HEAD (Skull).—Size? Shape? Symmetrical? Dolichocephalic? Brachycephalic? Fontanelles? Hydrocephalus? Bulging (Frontal or occipital)? Position of head? Stigmata of Degeneration (such as prognathism, de-

formity of ears, hair-lip)? Pain on percussion of skull? Mental condition? Speech? Sense of smell? (use peppermint).

EYES (Fundus).—Vision? Field of vision? Pupils? Light and accommodation reflexes? Ocular movements? Nystagmus (Lateral, vertical, or rotatory)? Is the cornea sensitive? Photophobia? Hearing?

FACE.—Symmetrical? Paralysis? Tongue? Taste? Deglutition? Articulation? Sensation in face? Teeth?

UPPER EXTREMITIES.—Are they equally developed? Position? Circumference of arm and forearm? Movements (of shoulder, arm, forearm, wrist, fingers)? Paralysis? Tonus of muscles? Are muscles atrophied or hypertrophied? Reflexes? Contractures? Electrical reactions? Sensation?

SPINE.—Rigidity? Lateral curvature? Kyphosis? Lordosis? Pain on percussion?

TRUNK.—Respiration? Sensation? Reflexes (Abdominal, Epigastric, Cremasteric)? Action of muscles?

LOWER EXTREMITIES.—Are they symmetrical? Circumference of thighs and calves? Ability to stand? Romberg's symptom? Ability to walk? Gait (Paretic, Spastic-paretic, Ataxic, Cerebellar)? Movements of individual groups of muscles? Is child able to raise thigh? To flex and extend thighs, legs, toes? To stand on tip-toes? To elevate toes, keeping heels on ground? Are muscles paretic or paralyzed, atrophied or hypertrophied? Electrical reactions? Reflexes (Knee-jerks, Ankle clonus, Babinski reflex, plantar)? Contractures? Sensation?

VESICAL AND RECTAL REFLEXES?

Examination of the head of a child often gives us valuable information. The normal head should be well rounded and symmetrical. According to the age of the child the size will vary. The average horizontal circumference at birth (measured by a line passing from the glabella around the occipital protuberance) is between 38 and 42 cm.; at the end of one year between 45 and 52 cm., and in later years it may grow gradually to 56 cm. Any marked departure from these measurements is abnormal, but heads of tolerable size may be associated with deficient development of parts of the brain. I have seen cases with normal circumference in which the anterior defect was not evident in the measurement in consequence of a slightly excessive development of the occiput. A normal circumference is also present at times, although the actual cranial capacity may be very much diminished by a receding frontal bone.

The following table will give the chief measurements of the skull in children; some allowance should be made for the thickness of hair and scalp.

TABLE OF CRANIAL MEASUREMENTS IN CHILDREN.

	New-born.		End of 1st yr.		1st to 7th yr.		10th year.		
	M.	F.	M.	F.	M.	F.	M.	F.	
1. Circumference.	34.0	34.0	42.0	42.0	34 to 46	....	49	47	Taken around glabella and occipital protuberance.
2. Binauricular arc...	20.0	20.0	25.5	25.0	.....	27	.....	....	Measured from B to opposite ext. aud. meatus.
3. Volume ...	385 to 450	....	700 to 1,000	....	.....	....	1,300	....	Volume is to circumference as 1,350 is to 50 (in the adult).
4. Naso-occipital arc.	20.0	22.0	28.0	28.0	.....	....	.....	....	N, β, T.
5. Naso-bregmatic arc.	7.7	7.7	.....	10.0	.....	....	.....	12	N, β.
6 Bregmatic lamb. arc.	9.0	9.0	.....	10.0	.....	....	12	....	β to A.

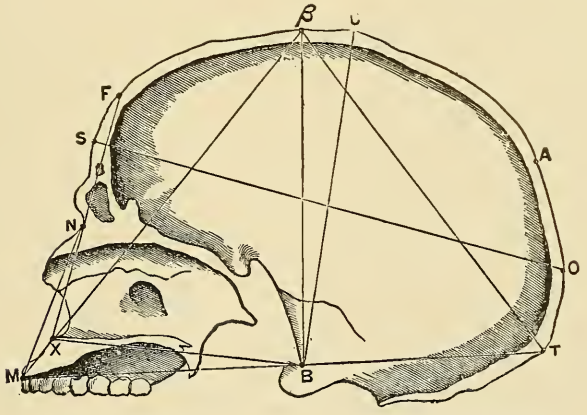


FIG. 1.—Craniometrical Lines. (Benedict and Peterson.)

The formula for the cephalic index is length : breadth :: 100 : x. An index below 78 is dolichocephalic ; 78 to 80 mesocephalic ; above 80 brachycephalic. The facial length is determined by a line passing from N to lowest part of chin.

Both halves of the head should be symmetrical. Asymmetry occurs chiefly in connection with defective develop-

ment of the brain and with early cerebral lesions. (Fig. 2.) The chief abnormalities of skull formation are as follows: Dolichocephalus, a long skull, the anterior posterior diameter being proportionately greater than the transverse. Many newborn children are dolichocephalic as the result of compression of the head in the pelvic canal, but after a few days or weeks the head is well rounded. Brachycephalic—the skull is short in the antero-posterior diameter. The terms microcephalus and macrocephalus need no further explanation.*

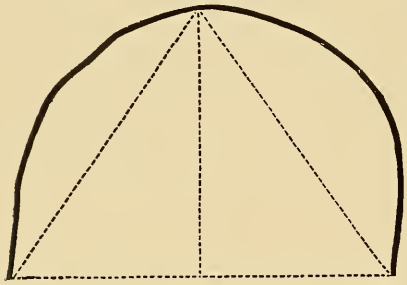


FIG. 2.—Asymmetry of Skull in a Male, aged Six Years. Right Hemiplegia from Birth. (Peterson.)

Bulging of the frontal or occipital bones is important as an indication of hydrocephalus. If there is a very considerable increase of intracranial fluid the sutures may be pushed asunder and can be felt distinctly through the scalp. This same condition occurs in some cases of neoplasm. In passing the hand over the head the fontanelles can be felt. The occipital fontanelle should be closed after a few weeks, the anterior remains open until the tenth or twelfth month. If it closes long before this period there is premature ossification of the sutures; if it remains open much longer, it is a probable sign of rickets.

After the examination of the head we may pass at once to the inquiry into the mental condition of the child. According to its age we must ask whether it recognizes its mother or nurse; whether it has learned to play, to understand what is said to it; whether it begins to imitate sounds, to articulate, etc.; in short, whether it shows a normal awakening of the mind. In children a little more advanced in years it is necessary to determine whether the child is able to keep up with others of its age; whether it

* A few special terms have been in use for oddly shaped heads: Keel-shaped skull, scaphocephalus; triangular skull, trigonocephalus; steeple-shaped skull, oxycephalus; obliquely deformed skull, plagiocephalus.

has been able to acquire the ordinary rudiments of knowledge. The physician should assure himself of these points by a personal examination, and should not depend upon the statements of parents and relatives.

A few extracts from Preyer's observations on his own child will show what may be expected of a normal child at different stages of its development:

DURING FIRST MONTH.—Recognizes difference between light and dark objects (even on first day); follows with its eyes object moved slowly before it (as early as eleventh day); begins to hear about the fourth day; recognizes sounds toward end of first month; learns to distinguish between bitter and sweet; recognizes disagreeable odors; first tears on twenty-third day during a crying spell; expresses displeasure by turning head away, by shutting its eyes, and, of course, by crying; begins to smile.

DURING SECOND MONTH.—Recognizes human voices and direction from which sound comes; turns head toward low sounds; is quieted by song; smiles when music is heard; recognizes its mother.

DURING THIRD MONTH.—Moves arms, expressive of pleasure; listens attentively; is able to support head a little; uses definite sounds in crying.

DURING FOURTH MONTH.—Associated eye movements perfect; stares at new objects; recognizes strange surroundings; reaches after distant objects; first attempt to sit upright.

DURING FIFTH MONTH.—Recognizes strangers as such; likes to take hold of everything; stretches out its arms to be taken up; holds head straight; sits alone; moves legs as if to walk; forms syllables.

DURING SIXTH MONTH.—Distinguishes faces; stares at strangers; smiles, if smiled at; smiles with relatives, not with strangers; turns its head toward a person leaving the room; begins to creep; "crows."

DURING SEVENTH MONTH.—Follows objects dropping out of its hands; recognizes its image in mirror with evident pleasure; points with finger at pictures; purposive movements; associates persons and names; extends hand when asked; articulates a number of different sounds in crying and in "lolling to itself."

DURING EIGHTH MONTH.—Sits upright when it is carried; some children attempt to stand and to walk.

DURING NINTH MONTH.—Begins to imitate tunes; laughs heartily; begins to beg for things.

DURING TENTH MONTH.—Takes an intense interest in its food; recognizes parent after absence of several days; he begins to walk alone; answers questions by motions and indicates where certain things are.

DURING ELEVENTH MONTH.—Stands quite alone; pushes chairs; makes first attempt to repeat sounds impressed upon its mind; begins to articulate its own name; understands language fairly well.

DURING TWELFTH MONTH.—Imitates laughter of others; stretches its

arms out to enforce its demands; improvement in walking and standing; looks at others attentively while they eat.

During Fourteenth, Fifteenth, and Sixteenth Months.—Independent speech is acquired, and repeats spoken words easily; in seventeenth month may speak short sentences, using verbs; from this time on there is steady improvement in memory of words and use of language.

At two years child may learn to repeat rhymes, to detect colors, etc.

If the mental condition of the child has been satisfactorily determined, the special senses should be examined. "Does the child see?" is a question often answered affirmatively by the mother, when a closer examination proves that the child is totally blind. Mothers are easily deceived in this, for the restless movements of the eyes in young children are supposed to be purposive and part of the visual act. To test vision use a candle or a taper, and pass it in front of the eyes at some distance from the head, so as to avoid heat sensations, and note whether the child follows the light. Do not be misled by accidental movements and avoid using rattles, for the child may turn its eyes in

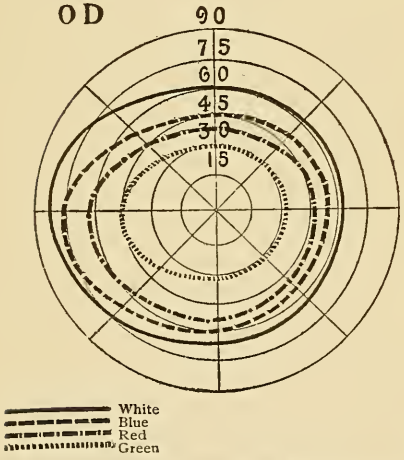


Fig. 3.—Normal Visual Fields for Colors.

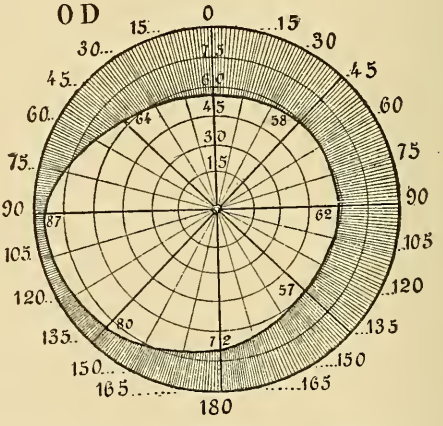


Fig. 4.—Normal Visual Field. (Grasset et Rauzier.)

the direction from which the sound issues without seeing the object. Use also simple substances of different colors. A blind child will not notice an approaching finger until the eyelashes have been touched.

It is a matter of still greater difficulty to test the field

of vision. In very young children it may be altogether impossible, but after a child has reached the age of five months or thereabouts, it may be possible to make a rough test of the visual field by passing objects from above and below, as well as from the sides, into the visual field, and noticing when the child begins to turn its eyes toward this

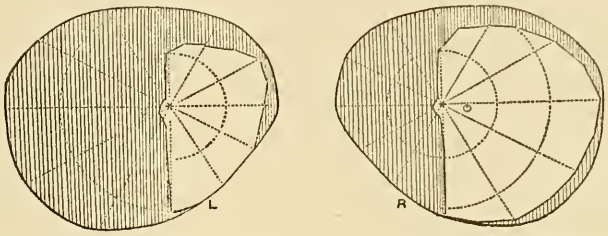


FIG. 5.—Field of Vision in a Case of Left-sided Hemianopsia. The Shading Represents the Blind Part; the Oval Outline is the Average Normal Field. (Gowers.)

object. The ordinary test as applied in the adult cannot be employed for children until they have developed considerable intelligence.

The condition of the visual field is often overlooked and the defects referred to are much more frequent than they are generally supposed to be. An examination should be made in all cases of cerebral palsies, and in those in which cerebral tumor is suspected.

In cases in which a very complex examination seems desirable, and these are relatively few, special tests should be made for form and color. The hemianopsic defects as they occur in tumors, and in cerebral palsies (first described by Freud), the concentric limitations occurring in hysteria, are the most common defects in children.

The pupils should be equal and contract promptly to light and during accommodation. They should also contract if the patient attempts to close the eyes while the examiner endeavors to keep the lids open (A. Westphal and Piltz).

The ocular palsies will be best understood by reference to the subjoined table. Conjugate deviation is due to disease in the hemisphere, and according to Grasset to lesions in the first and second frontal convolutions and in the angular gyrus and its vicinity; the patient looks toward the lesion except in the case of spasm of the muscles, when the patient looks away from the lesion. Nystagmus occurs in many cerebral affections, particularly in cases of early cerebral disease or congenital defect. It is seen also in cases in

which there are stigmata of degeneration as well as in multiple sclerosis. In the latter it may be bilateral or rotatory. The corneal reflexes (the prompt closure of the eyelids if the cornea is touched carefully with the head of a pin or a small lead-pencil) should be tested. In a great many individuals this reflex does not ensue upon irritation of the conjunctiva, a fact that has deceived many physicians.

MUSCLES OF THE EYES AND FACE.

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Ciliary ...	Makes lense more convex; aids in accommodation.	Loss of accommodation; spasm of ciliary muscle may cause myopia.	The third nerve.	Corpora quadrigemina and peduncle.	Meningitis; traumatism to skull and eye; diphtheritic palsy; diseases of mid-brain; tumor; polioencephalitis superior; in cerebral syphilis; hereditary ataxy; poisoning by drugs.
Sphincter iridis.	Contraction of pupil to light and during accommodation.	Paralytic mydriasis; no contraction to light and during convergence or accommodation.	The third nerve.	Corpora quadrigemina and peduncle.	
Dilatator pupillæ.	Dilates pupils; as a result of sensory or psychic stimulation.	Pupil does not respond to sensory stimulation.	Sympathetic		
Rectus superior.	Moves eye upward and inward. Acts with inferior oblique.	Upward movement limited; diplopia; false image above; deficient rotation of eyeball.	The third nerve.	Corpora quadrigemina and peduncle.	Diseases in mid-brain region; tumor; polioencephalitis superior; meningitis; cerebral syphilis; multiple neuritis (particularly after diphtheria); congenital nuclear disease.
Rectus internus.	Moves eye inward.	Strabismus divergens; defective inward pull.	As above.	Corpora quadrigemina and peduncle.	
Rectus inferior.	Moves eye downward and rotates it inward. Acts with superior oblique.	Imperfect movement downward; eye rotated outward.	As above.	Corpora quadrigemina and peduncle.	
Obliquus inferior.	Acts with rectus superior, moving eye upward and outward, and rotates it upward.	Imperfect movement upward; eye rotated inward.	As above.	Corpora quadrigemina and peduncle.	
Obliquus superior.	Moves eye downward and outward. Acts with inferior rectus; rotates downward.	On looking downward eye is pulled inward; convergent strabismus; diplopia on stepping downward.	The fourth nerve (trochlearis).	Peduncle near the corpora quadrigemina.	

MUSCLES OF THE EYES AND FACE.—*Continued.*

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Rectus externus.	Moves eye outward.	Outward movement impaired; head turned in direction of paralyzed muscle.	The sixth nerve (abducens).	Pons	As before, and in disease of the pons.
Levator palpebræ superioris	Raises upper eyelid.	Ptosis; eye closed; may be opened a little by frontalis muscle.	The third nerve.	Peduncle and corpora quadrigemina.	Associated with other third nerve diseases; often congenital.
Orbicularis palpebrarum.	Closes eyelids.	Eyes cannot be closed (lagophthalmos).	The seventh nerve.	Pons	Peripheral facial neuritis; affections at base of brain (meningitis, tumor, and the like); in some dystrophies.
Frontalis and corrugator supercilii	Raises eyebrows; folding of skin of forehead, as in frowning.	Imperfect raising of eyebrows; no frowning; lines of forehead disappear.	As above.	As above.	As above.
Orbicularis oris, buccinator, and other muscles of face.	Move lips and cheeks, as in speaking, kissing, showing of teeth, and the like.	Face distorted and pulled toward healthy side; inability to pout lips or to whistle; dribbling of saliva from paralyzed side; flapping of cheek with each expiration, owing to paralysis of buccinator; disappearance of nasolabial fold.	The seventh nerve.	Pons	Cerebral apoplexies (escape of upper branches of facial); peripheral facial palsy (often due to exposure); in all lesions of pons; in dystrophies.
Masseter, temporal, and pterygoids.	Masseter, temporal, and internal pterygoid elevate lower jaw; external pterygoid draws jaw forward; the two pterygoid muscles of one side acting together move teeth toward opposite side.	Mastication imperfect; if in spasm, jaws cannot be opened; jaw moved toward paralyzed side by action of pterygoids.	The fifth nerve.	Pons	Rarely affected in children except in tetanus; in facial hemiatrophy; in lesions of pons.

Mydriasis can be caused by paralysis of sphincter iridis (third nerve), or by spasm of the dilator muscles (mydriasis spastica). Atropin, Duboisin, and Hyoscyamia have a similar effect by paralyzing the sphincter, and con-

tracting the dilator. Cocaine produces a transitory mydriasis by irritation of the pupillary fibres of the sympathetic.

Myosis may be due to irritation of the sphincter supplied by the third nerve, and occurs in early stages of many cerebral affections, including apoplexy; or it may be due to paralysis of the dilator muscle.

Inequality of pupils is sometimes congenital, more often due, however, to cerebro-spinal disease (meningitis, syphilis, multiple sclerosis, hereditary ataxia). Muscles of both eyes act conjointly. If such action is deficient we speak of conjugated paralysis. There may be paralysis of lateral, upward or downward movements, without total palsy of any one muscle. If individual muscles are paralyzed, or muscles in one eye only, double vision is certain to result.

In diplopia the exact position of true and false images will help to determine the character of the ocular palsy. If one muscle is paralyzed secondary deviation of the associated muscle in the other eye may take place.

Paralysis of the inner muscles of the eye is designated as ophthalmoplegia interna; paralysis of the muscles moving the eyeball, as ophthalmoplegia externa, and such ophthalmoplegia may be total or partial. Total ophthalmoplegia externa and interna is the occasional accompaniment of disease at the base; partial ophthalmoplegia (only part of the muscles supplied by the third nerve, for instance, being affected) points to nuclear disease, but syphilitic infiltration of the root fibres may simulate nuclear disease.

Narrowing of the orbital fissure (sometimes congenital) may be due to paralysis of smooth fibres in the lid, innervated by the sympathetic. Clonic movements of eye muscles we term nystagmus; this may be lateral, vertical, or rotatory.

An examination of the sense of hearing is often called for. In children of defective development this sense may be entirely wanting. The simplest tests are to clap the hands at a distance from the child's head, to whisper its name, to use a loudly ticking watch—even very young children will be attracted by the sound if it is perceived. In the case of older children tuning-forks may be applied to the head. In disease of the ear proper the vibrations would be perceived, but not so if the nerve itself, the labyrinth, or the auditory tract were involved. Hyperacusis occurs in amaurotic family idiocy, in some forms of meningitis.

Asymmetry of the face may point to congenital defect or to a preceding palsy. It is of some interest to know that asymmetry of the face and of the nose, adhesion of the ear-lap, malformations of the outer ear, the existence of a torus palatinus (felt as a ridge in the roof of the mouth) are the ordinary stigmata of a degenerative type, of which so much has been made by the Italian criminologist of the

present day. If such stigmata prove a predisposition to the development of nervous troubles, to crime, or to insanity, it is well that the physician should know of them.

The teeth play a very important part in the life of every child. If not developed between the ages of six months and one year, rickets must be suspected. Notched teeth (second dentition) are suspicious of syphilis. Unusually early development of the teeth is an evidence of premature ossification.

MUSCLES OF TONGUE, PALATE, AND PHARYNX.

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Genio-glossus.	Pushes tongue to opposite side.	Tongue when protruded deviates to paralyzed side.	The twelfth nerve (hypoglossal).	Medulla....	
Styloglossus.	Raises tongue backward and upward.	Tongue cannot be moved backward or hollowed out (action deficient in many healthy subjects).	The twelfth nerve.	Medulla....	Bulbar palsies (acute and chronic); in specific and tubercular diseases of base; dystrophies (rare).
Lingual muscle proper.	All movements of the tongue itself.	When lying in mouth deviation to healthy side; when protruded deviates to paralyzed side; if one or both halves are atrophied tongue looks shrivelled.	The twelfth nerve.	Medulla....	
Azygos uvulæ.	Shortening of uvula.	Uvula deviates toward sound side; if both sides are paralyzed there is nasal tone and regurgitation through nose.	Probably pharyngeal plexus; seventh nerve (?).	Medulla....	As above
Levator palati.	Raises the velum palati.	Arch cannot be raised in the intonation of "ah;" if paralysis is bilateral flapping of arch and regurgitation of food through nose.	As above.	Medulla....	As above; see also seventh nerve affections.
Palato-pharyngeal muscles.	Prevent food from passing toward upper part of pharynx and posterior nares.	Regurgitation of food; nasal speech.	The fifth nerve.	Pons.....	Basilar affections.
Stylo-pharyngeus.	Helps to draw larynx upward so as to be closed by epiglottis and overtopped by tongue.	Imperfect deglutition; food gets into windpipe.	Glosso-pharyngeal.	Medulla....	Bulbar affections and diseases of the base.

MUSCLES OF TONGUE, PALATE, AND PHARYNX.—*Continued.*

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Constrictors of pharynx.	Help to push food into gullet.	Food is swallowed very imperfectly; sticks in throat.	Pharyngeal plexus.	Medulla....	Diseases of the base (bulbar).
Laryngeal muscles.	Movements of vocal cords in respiration and in articulation.	Hoarseness and difficulty in breathing; laryngoscopic examination reveals false position of vocal cords (see special text-books).	Recurrent laryngeal nerve excepting the crico-thyroid muscle.	Medulla....	Bulbar troubles (similar symptoms may be caused by tumors and foreign bodies in larynx).

The movements of the tongue have been considered in the annexed table. A short frenulum may cause difficulty in articulation, but it is often suspected to be the cause in cases in which the defect is due to a distinct cerebral lesion.

Sensory disturbances of the face are rare in children, but, if suspected, tests should be made carefully with cotton, with the head and point of a pin, and by application of hot and cold objects. Subjective sensory disturbances (neuralgia) may occur as in adults, and will vary in distribution according to the branches affected. (Fig. 6.)

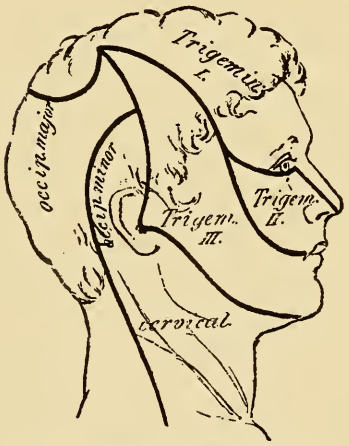


FIG. 6.—Sensory Chart of Face. (C. S. Freund.)

In examining the trunk and the four extremities no fact is more important to establish than the existence of paresis, or paralysis of individual muscles, or of groups of muscles. To be able to do this the examiner must know the action of all the more important muscles in health and the disturbances due to disease of such muscles. These are the very facts in which the student's knowledge is, as a rule, most defective. In the appended tables I shall endeavor to give the principal points in concise fashion:

MUSCLES OF HEAD AND NECK.

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Sterno-cleido-mastoid.	Raises and turns face to opposite side; head inclines to same side; if both muscles act conjointly head is brought forward.	Inability to raise head from bed, or other horizontal position, if both muscles are affected; if one muscle is affected, no marked change of position, unless opposite muscle is contractured; spasm of muscle frequent; head inclined to one side.	Spinal accessory.	Medulla and second and third cervical segments.	In bulbar and cervical cord affections; in later stages of progressive muscular atrophies; occasionally in neuritis.
Rectus capitis anticus major.	To flex head.	Cannot flex head so as to bring chin on chest.	Upper cervical.	Upper cervical segments.	Diseases of the cervical region (myelitis, meningitis, tumor; progressive wasting of muscles.
Rectus capitis anticus minor.	To flex head.				
Rectus capitis lateralis.	Slight rotation.	Deficient rotation scarcely noticeable, unless sterno-cleido-mastoids are diseased.			
Scaleni anterior medius, et posterior.	Elevate ribs when vertebral column is fixed; aid in inspiration; slight lateral flexion.	Deficient inspiratory movements.	Lower cervical nerves.	Lower cervical segments.	
Longus colli.	Flexor of vertebral column.	Imperfect flexion of upper spine.	Lower cervical nerves.		

The sterno-cleido-mastoid and the other muscles of the head and neck, mentioned in these two tables, are more frequently the seat of spasm than of paralysis. The spasm may be restricted to one muscle or may involve several. It may be the result of organic disease in the medulla oblongata or spinal cord. More often it is functional in character and related to other spasmodic conditions. The same group of muscles may be involved in the ordinary rheumatic affections (torticollis, caput obstipum).

MUSCLES OF SHOULDERS AND UPPER EXTREMITY.

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Trapezius. 1. Clavicular portion (respiratory; outer third of clavicle to occipital bone).	Pulls head backward; rotates slightly toward side of muscle, so that chin is turned to opposite side; contraction of both clavicular portions, bends head backward; slight elevation of shoulders; aids in deep inspiration.	Deficient backward movement of head; not marked as rule because deep muscles perform this function; shoulder does not move during inspiration.	Spinal accessory.	Medulla and second and third cervical segments.	Progressive muscular wasting; diseases of medulla and upper cervical cord; clavicular portion least frequently involved.
2. Middle portion (from acromion and outer spine of scapula to ligament. nuchæ and upper dorsal spines).	Raises shoulder-blade; elevation of acromion (clavicle goes along).	Acromion depressed by weight of upper extremity; inner upper angle may be pulled upward by levator anguli scapulæ; internal lower angle is nearer to median line.	Spinal accessory nerve.	As above.	As above.
3. Lower portion and adductor.	Adduction of scapula toward median line.	Margin of scapula is about ten ctm. distant, instead of being five or six ctm. distant from median line; loss of adductor may be covered up by action of rhomboids; rounding of back. (Fig. 8.)	Spinal accessory nerve.	Medulla and second and third cervical segments.	
Rhomboids.	Oblique movement of scapula from below, upward and inward, so that inferior angle is brought nearer the median line; hold spinal margin of scapula down to thorax.	Deep groove between inner margin of scapula and thorax; if serratus is normal, this groove disappears if arm is extended forward; shoulder-blade cannot be approximated to median line. (According to Duchenne this can be affected by upper portion of latissimus dorsi.)	Fifth cervical.	Fourth and Fifth cervical segments.	As above.

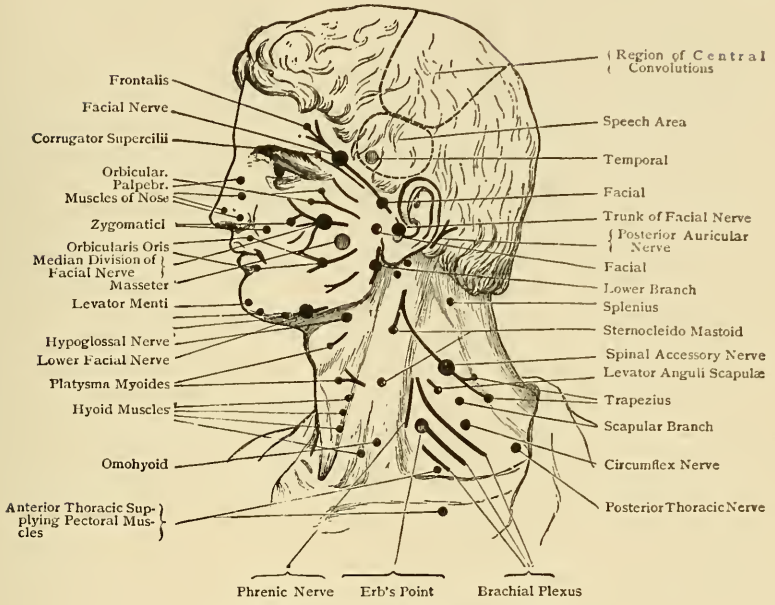


FIG. 7.—Motor Points of Face. (Erb.)

FIG. 8.—Patient with Hypertrophy of Infra and Supra Spinati, Showing Rotation of Right Scapula and Deep Groove along Inner Margin of Scapula (Atrophy of Rhomboids and Slight Atrophy of Lower Portion of Trapezius).

MUSCLES OF SHOULDERS AND UPPER EXTREMITY.—*Continued.*

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Levator anguli scapulæ.	Draws superior inner angle of scapula upward: aids in shrugging of shoulders.	Isolated paralysis rare.	Third and Fifth cervical nerves.	Second and Fourth (?) cervical segments.	Dystrophies and cervical diseases.
Serratus magnus.	Rotation of shoulder-blade outward, and slight elevation of acromion; holds inner margin of scapula to thorax; brings arm from horizontal to vertical position.	Scapula pulled upward; lower inner angle nearer the median line; arm cannot be raised above horizontal position; if arm is stretched forward scapula is removed from thorax ("winged scapula"); during abduction of arm, scapula is moved nearer to median line, and crowds trapezius and rhomboids forward.	Posterior thoracic nerve.	Fifth and sixth cervical segments.	Progressive muscular atrophies (dystrophies); neuritis of part of the brachial plexus; after traumatic injuries to shoulder; in cervical cord affections.
Deltoid (three divisions).	To raise arm to horizontal position, and forward, outward, or backward; movements possible only if scapula is fixed by action of serratus and trapezius.	Can raise shoulder but not arm; shoulder flattened (atrophy); groove between acromion and head of humerus; each division of deltoid may be paralyzed singly.	Circumflex.	Fourth, fifth, and sixth cervical segments.	As above; also in Erb's form of obstetrical paralysis.
Infraspinatus. Teres minor.	Rotator humeri posticus (Duchenne); rotate arm outward.	Arm cannot be moved outward. Difficulty in writing (Duchenne).	Supra-scapular. Circumflex.	Fourth, fifth, and sixth cervical segments.	As in case of deltoid.
Subscapularis.	Rotator humeri anticus (Duchenne); rotates arm inward.	Arm cannot be moved inward; scapula is rubbed against ribs.	Subscapular nerve.		
Supraspinatus.	Helps to steady shoulder-joint and to elevate arm forward and outward; outer angle of scapula is depressed.	According to Duchenne, humerus is separated still further from acromion. if supraspinatus is affected in addition to deltoid.	Suprascapular.	Fourth cervical.	As above.

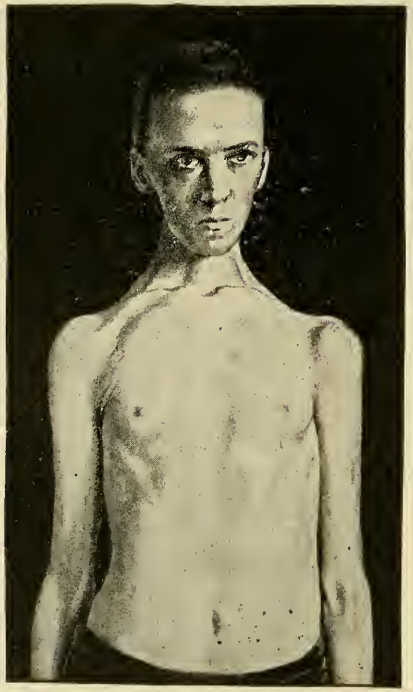


FIG. 9.—Case of Progressive Muscular Dystrophy, Showing Atrophy of Trapezii and Abnormal Position of Clavicles.

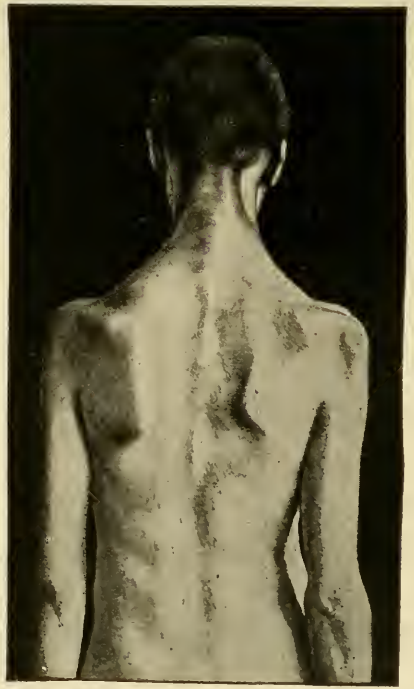


FIG. 10.—Same Patient; Showing Particularly the Atrophy of the Trapezii and the Scapular Muscles; Head Inclined Forward; Change in Position of Scapulæ; Inferior Angle of Right Scapula Nearer the Median Lines.

MUSCLES OF SHOULDERS AND UPPER EXTREMITY.—*Continued.*

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Latissimus dorsi.	Pulls the arm, when raised, downward and backward; if arm is at rest upper portion brings scapula nearer the median line; united action of upper third of both muscles causes extension of dorsal trunk; single action causes lateral movement of trunk.	Arm cannot be moved backward; insufficient extension of dorsal spine; trunk cannot be moved laterally.	Subscapular, also branches of dorsal and lumbar nerves passing through muscle.	Sixth and seventh cervical.	As in progressive atrophies and dystrophies; in cervico-dorsal lesions; in neuritis.
Teres major.	Rotates raised humerus inward; adduction of arm to thorax; slight elevation of shoulder.	Very few symptoms; action supplied by other muscles.	Subscapular.	Seventh cervical.	As above.
Pectoralis major.	Clavicular portion depresses humerus from raised position to horizontal; adduction of arm, as in giving a blessing; sternal portion depresses arm completely, and if arm is at rest draws acromion forward and backward.	Imperfect adduction of arm; paralysis can be discovered best by extending arms and trying to press volar surfaces against each other.	Anterior thoracic.	Fifth, sixth, and seventh cervical.	Amyotrophies and dystrophies, chiefly; also in lesions of brachial plexus.

Absolute paralysis of both upper extremities is rare in children. If an entire extremity is paralyzed, the arm either hangs limp by the side of the body, or is flexed at the elbow. If the arm is lifted it falls by its own weight. Children sometimes refuse to make an effort, or do not understand what is wanted of them. After a few trials it is generally possible to decide whether there is a loss of power or mere lack of effort. Total paralysis of the upper extremity occurs either from disease of the brachial plexus, from spinal cord affections, or from cerebral lesions. If due to the first or second causes, the

FIG. 11.—Young Boy with Multiple Neuritis, showing Double "Wrist Drop" and Slight "Foot Drop."

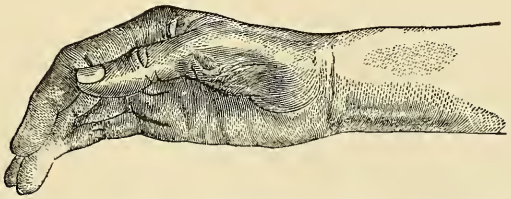


FIG. 12.—Appearance of Hand in an Early Stage of Progressive Muscular Atrophy; Atrophy of Abductor Brevis and Opponens Pollicis. (Duchenne.)

paralysis is flaccid; if due to the third cause, the paralysis is spastic, and very often part of a hemiplegia. Plexus lesions are observed in very young children (obstètrical palsies), spinal lesions in children of any age, and cerebral lesions in children under the age of four chiefly; but the after-effects of these conditions are often visible late in life. From the causes enumerated above, as well as in cases of neuritis and progressive muscular diseases, the paralysis may be incomplete and restricted to definite groups of muscles only. To test the extent of paralysis, the patient should be asked to perform the various movements, such as raising the shoulder, putting out arm, flexing and extending the elbow, wrist, and fingers. In very young children these tests cannot always be satisfactorily made, but even in infants much can be inferred from their power to hold or to grasp objects placed in front, above, or below them. In older children further tests can be made by offering resistance to active movements, and asking them to overcome it. The ability to bring the fingers in opposition to one another, to make the thumb touch the tips of the other fingers, the moving of the fingers to and from the middle one, and the ability to write, are necessary tests of the action of the intrinsic muscles of the hand.

MUSCLES OF ARM, FOREARM, AND HAND.

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Triceps...	Extends forearm; long head of triceps, and coraco-brachialis help to keep head of humerus in position.	Arm cannot be extended except by its own weight; if long head of triceps is affected subluxation of head of humerus occurs easily.	Musculo-spiral.	Sixth, seventh, eighth cervical segments.	Poliomyelitis and other affections of cervical cord; traumatic injuries; amyotrophies and dystrophies (triceps escapes in many peripheral palsies).
Biceps....	Flexion and supination of forearm.	Flexion deficient, but can be carried out in part by other muscles.	Musculo-cutaneous.	Fourth, fifth, sixth cervical.	
Supinator longus	Flexes forearm and aids in pronation.	Flexion and pronation deficient; muscle does not stand out prominently if arm is flexed and attempt is made by another to extend it forcibly; if muscle is atrophied arm is spindle-shaped	Musculo-spiral.	Fourth, fifth cervical.	As above; involved in peripheral neuritis (traumatic), not in lead palsy.
Supinator brevis.	Supinates hand when forearm is extended.	Deficient supination of hand.	Musculo-spiral.	Fifth cervical.	Diseases as above; also in peripheral palsies.

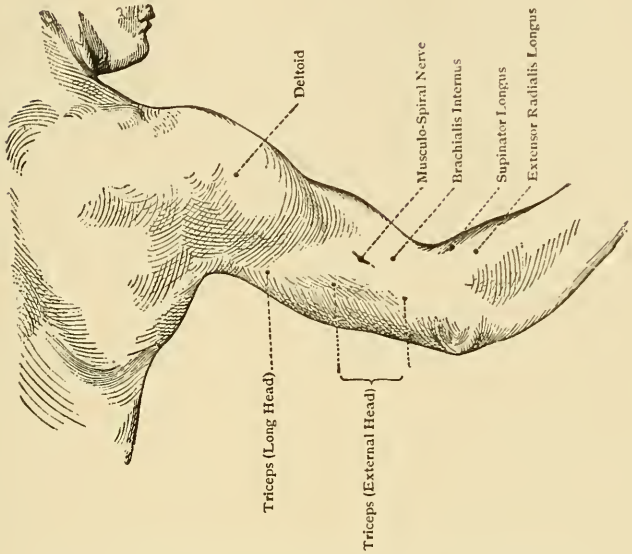


FIG. 13.—Motor Points on Shoulder and Arm. (Bernhardt.)

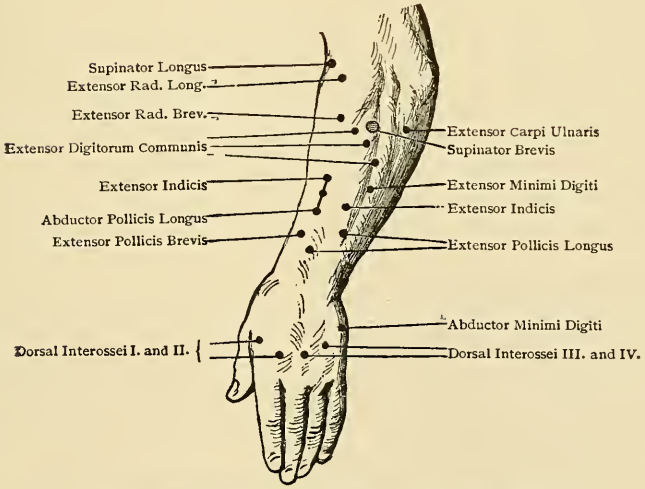


FIG. 14.—Motor Points of Forearm and Hand. (Erb.)

MUSCLES OF ARM, FOREARM, AND HAND.—*Continued.*

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Extensor carpi radialis longus et brevis.	Extension and abduction of wrist: the shorter muscle has pure extension action only.	Wrist cannot be flexed dorsally (extended) or abducted; flattening of forearm. (Fig. 11.)	Musculospiral.	Seventh cervical.	As before; especially in neuritis.
Extensor carpi ulnaris.	Extension and abduction of wrist.	Wrist cannot be flexed dorsally or adducted; "drop wrist" is characteristic of paralysis of extensors.	As above.	Seventh cervical.	As above.
Extensor digitorum communis. Extensor indicis. Extensor minimi digiti.	Extension of first phalanges of all fingers and abduction.	First phalanges cannot be extended nor fingers abducted; grasp is weak because flexor muscles are shortened and cannot contract forcibly.	Musculospiral.	Seventh cervical.	As above.
Flexor carpi radialis.	Flexion of wrist and pronation.	Deficient flexion.	Median.....	Eighth cervical.	As above.
Flexor carpi ulnaris.	Flexion of wrist and supination.	Flexion and supination impaired.	Ulnar.....	Eighth cervical.	As above.
Palmaris longus.	Flexion of wrist only.	Flexion impaired; no anomalous position of hand from paralysis of wrist as hand falls by its own weight; the flexors of fingers may act as substitutes.	Median.....	Eighth cervical.	As above.
Flexor digitorum sublimis.	Flexes second phalanx toward first.	Second phalanx cannot be flexed.	Median.....	Eighth cervical.	As above.
Flexor digitorum profundus.	Flexes last two phalanges toward first.	Last two phalanges cannot be flexed.	Ulnar and Median.	Eighth cervical.	As above; muscle should be tested with special care in cases of traumatic injuries.
Interossei and lumbricales.	Abduction and adduction of fingers if first phalanges are extended; flexion of first phalanges and simultaneous extension of second and third phalanges.	Fingers cannot be abducted or adducted; interosseous spaces are very marked; "Main en griffe" due to extension of first phalanges and flexion of second and third phalanges. (Figs. 17, 18.)	Ulnar, which also supplies third and fourth lumbricales; median supplies first two and sometimes third lumbricales.	Eighth cervical, first dorsal.	As above; often the first muscles to be affected in progressive spinal atrophies.

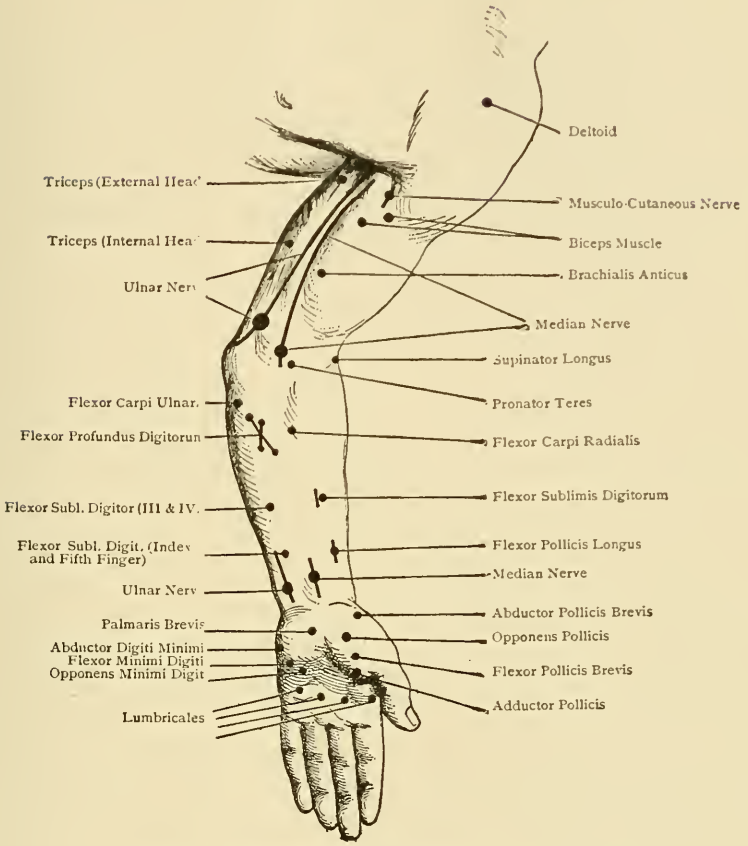


FIG. 15.—Motor Points on Inner Surface of Arm. (Erb.)

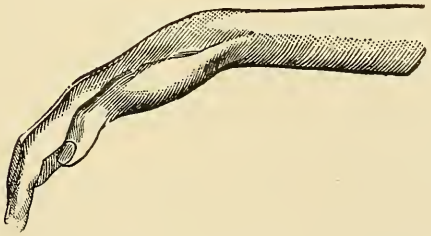


FIG. 16.—Extreme Atrophy of Thenar Muscles—"Ape Hand." (Duchenne.)

MUSCLES OF ARM, FOREARM, AND HAND.—*Continued.*

Name of Muscle.	Normal Function.	Symptoms of Deficient action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Thenar muscles: Extensor pollicis brevis.	Extends first phalanx and abducts metacarpal bone; acts with adductor pollicis longus.	Impairment of extension and adduction; flattening of ball of thumb.	Musculo-spiral.	First dorsal.	As before; more especially in amyotrophies and neuritis.
Extensor pollicis longus.	Extends both phalanges of thumb; also adduction of metacarpal bone and backward movement of thumb.	Deficient extension and adduction; second phalanx is flexed toward first.	Musculo-spiral.	First dorsal.	As above.
Abductor pollicis longus.	Abduction of metacarpal bone; aids in flexion of hand.	Deficient abduction of metacarpal bone; if this muscle and extensor pollicis brevis are paralyzed adduction results.	Musculo-spiral.	First dorsal.	As above.
Abductor pollicis brevis.	Opposition of thumb.	No opposition movement.	Musculo-spiral.		
Opponens pollicis and outer portion of the flexor brevis.			Median.	First dorsal.	As above.
Abductor pollicis brevis; flexor brevis and adductor.	Flex first phalanx and extend second phalanx (like interossei, also have an abduction and adduction action.	No flexion; if muscles are paralyzed and atrophied, ape hand is formed. (Fig. 16.)	Median and ulnar.		As above.
Flexor pollicis longus.	Flexes end phalanx.	No flexion of end phalanx.	Median.		As above.

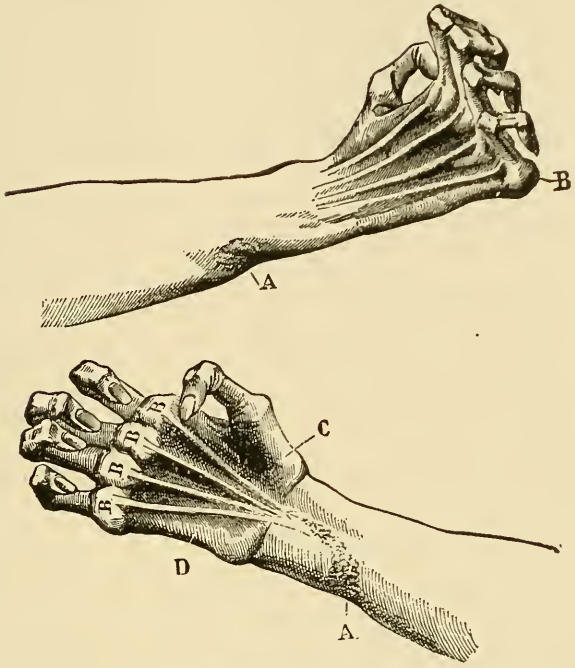


FIG. 17.—Extreme "Main en Griffe," after Injury to Ulnar Nerve A. Subluxation of Hypertrophied Heads of Metacarpal Bones B. (Duchenne.)

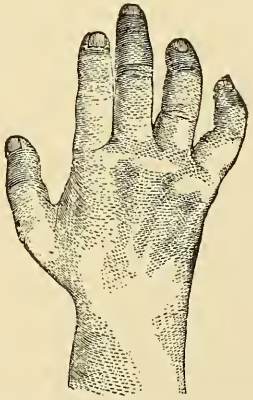


FIG. 18.—Slight Atrophy of Interossei, and Beginning of "Clawed Hand." (Duchenne.)

MUSCLES OF THE PELVIC GIRDLE AND LOWER EXTREMITIES.

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Gluteal muscles.	Gluteus maximus = extension of leg at hip and slight rotation outward.	No extension of thigh; great difficulty in climbing; no abduction of thigh; waddling gait, exaggerated movement of pelvis.	Inferior Gluteal (sacral plexus).	First and second sacral segments.	In progressive atrophies; these muscles are hypertrophied or atrophied in various forms of dystrophy; myelitis of lower cord; poliomyelitis; traumatic injuries to cord and cauda equina.
	Gluteus medius et minimus = abduction of leg; if leg is extended also lateral movement of trunk.		Gluteal superior.		
External rotators: Pyriformis. Gemelli. Quadratus femoris. Internal obturator.	Outward rotation of thigh.	Deficient outward rotation; leg turned inward.	Sacral plexus (muscular branches).	Fifth lumbar.	As above.
External obturator.			Obturator nerve (lumbar plexus)		
Ilio-psoas.	Flexion and outward rotation of thigh.	Flexion difficult; in bed thigh cannot be flexed; difficulty rising from horizontal position.	Crural (lumbar plexus)	Fourth lumbar.	
Tensor fasciæ latæ.	Flexion and inward rotation.		Superior gluteal.		As above.
Adductor muscles.	Adduction of thigh.	No adduction; thigh rolls outward.	Obturator nerve, great sciatic and crural.	Third lumbar.	
Sartorius.	Flexion of hip and knee, and slight outward rotation of thigh.	Flexion impaired; acts imperfectly.	Crural.	Third lumbar.	As above.
Quadriceps femoris.	Extensor of leg (rectus femoris also flexes hip).	Leg cannot be extended; to test it ask patient, who is lying down with hip bent, to stretch out the leg; when patient is sitting down to extend leg; or try to flex leg while patient extends forcibly in the sitting posture with leg hanging down.	Crural.	Fourth lumbar.	As above; very frequent in poliomyelitis.

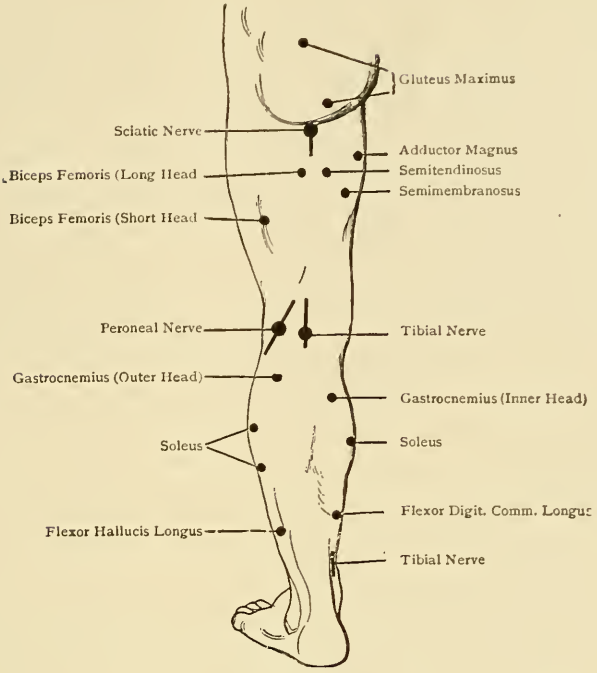

Gluteus Maximus
Sciatic Nerve
Adductor Magnus
Biceps Femoris (Long Head
Semitendinosus
Semimembranosus
Biceps Femoris (Short Head
Peroneal Nerve
Tibial Nerve
Gastrocnemius (Outer Head)
Gastrocnemius (Inner Head)
Soleus
Soleus
Flexor Digit. Comm. Longus
Flexor Hallucis Longus
Tibial Nerve

FIG. 19.

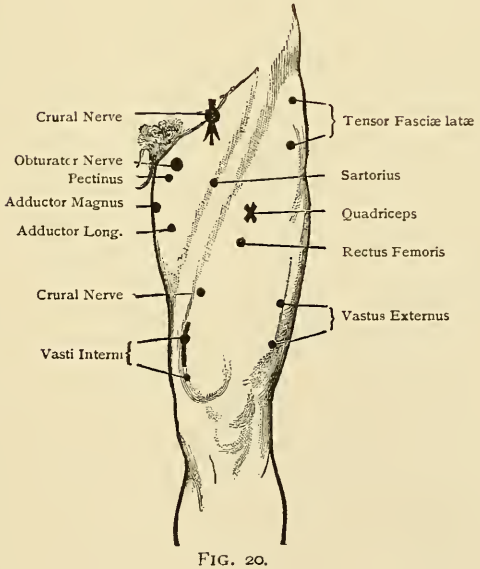

Crural Nerve
Tensor Fasciæ latæ
Obturator Nerve
Pectinus
Sartorius
Adductor Magnus
Quadriceps
Adductor Long.
Rectus Femoris
Crural Nerve
Vastus Externus
Vasti Interni

FIG. 20.

MUSCLES OF THE PELVIC GIRDLE AND LOWER EXTREMITIES.—*Continued.*

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Biceps; semitendinosus and semimembranosus.	Flexors of leg, and extensors of hip in ordinary walking, not in climbing stairs.	Deficient flexion; action of quadriceps may cause excessive extension; in standing; thigh is flexed to excess; trunk moved backward.	Sciatic.	Fifth lumbar.	As before.
Gastrocnemius (also plantarius and soleus).	Flexion and adduction of foot; toes point inward; first phalanges are extended; last phalanges in plantar flexion.	Deficient flexion of foot; heel cannot be raised; cannot stand on tiptoes.	Internal popliteal.	Fifth lumbar.	Most prominently affected in dystrophies and in poliomyelitis.
Peroneus longus.	Slight flexion, chiefly abduction of foot; elevates outer margin of foot.	Deficient abduction; plantar arch lessened; increased by contracture. (Figs. 21–24); flat-foot; walking tiresome.	Peroneal.	First and second sacral.	
Anterior tibial muscles (tibialis anticus, extensor digitorum, and extensor pollicis longus).	Extensors of foot and toes; tibialis anticus is also adductor; extensor digitorum in addition to moving toes, also elevates outer ridge of foot and abducts foot.	Deficient extension; "drop-foot," toes scrape floor; to clear this, excessive flexion at knee and hip; contracture of flexors and pes equinus or equinovarus. (Figs. 11, 26, 27.)	Anterior tibial.	Fifth lumbar, first sacral.	As above, but particularly in poliomyelitis; peripheral neuritis; frequent also in peroneal form of progressive muscular atrophy.
Posterior tibial muscle.	Adduction of foot; outer margin of foot becomes convex.	Deficient abduction or adduction; deformities result from deficiencies.	Posterior tibial nerve.	First and second sacral.	As above.
Peroneus brevis.	Abduction of foot.		Peroneal.		
Interossei pedis et lumbricales.	Abduction and adduction of toes; flexors of first phalanx; extend second and third phalanx.	Abduction and adduction of toes deficient; paralysis of interossei; hyperextension of first phalanges; second and third flexed (clawed foot). (Fig. 25.)	Posterior tibial.		
Adductor; flexor brevis and abductor hallucis.	Flex first phalanx of big toe and extend second.	Deficient flexion of toes; foot cannot be pushed off ground easily.	Posterior tibial.	First and second sacral.	

FIG. 21.—Submetatarsal Prominence. A Lower than Normal, and Increased Plantar Arch, resulting from Contracture of Peroneus Longus.

FIG. 22.—Exhibiting, in Addition, Prominence of the Tendon of the Peroneus Longus F. (Duchenne.)

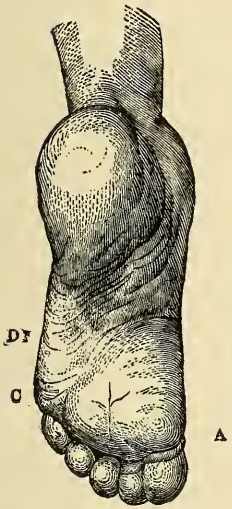


FIG. 23.—Plantar Surface of Same Foot, exhibiting Changes due to Contracture of Peroneus Longus, shortening of Transverse Diameter A C, and Torsion of Foot.

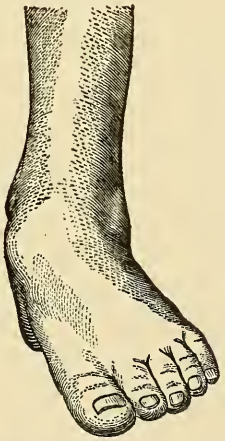


FIG. 24.—Same Foot. Valgus Position due to Same Cause.

FIG. 25.—"Clawed Foot" from Atrophy of Interossei and Other Intrinsic Muscles of Foot. (Duchenne.)

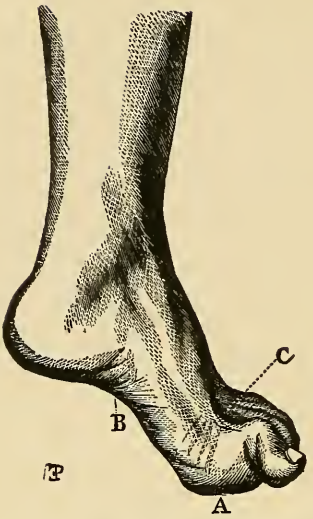


FIG. 26. — Pes Equinus in a Boy Five Years of Age from Atrophy of Tibialis Anticus.

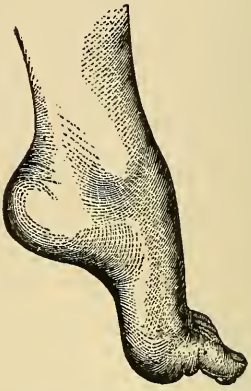


FIG. 27. — Pes Equinus of Eight Years Duration.

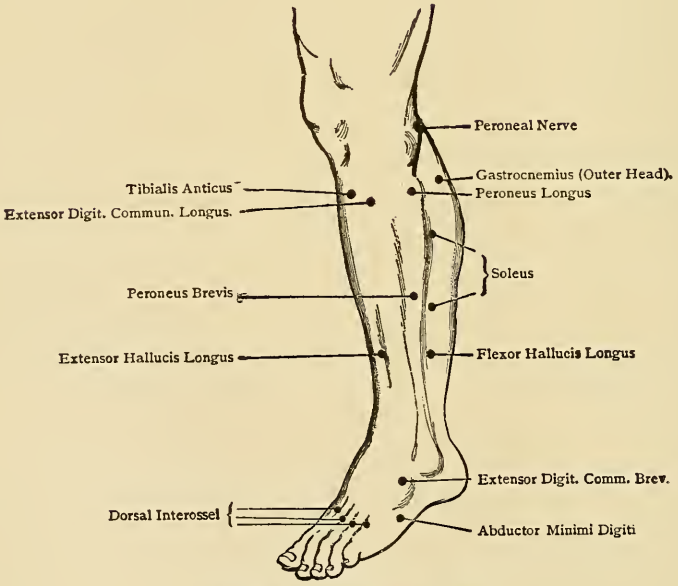


FIG. 28.—Motor Points. (Erb.)

MUSCLES OF BACK AND TRUNK.

Name of Muscle.	Normal Function.	Symptoms of Deficient Action.	Innervated by	Represented in	Diseases in which Muscle is commonly Involved.
Erector spinæ: sacro-lumbalis; longissimus dorsi.	Extension of lower dorsal and lumbar vertebræ.	Lordosis of lower spine; perpendicular line from shoulder falls behind os sacrum; unilateral palsy causes deflection of spine toward sound side. (Fig. 30.)	Dorsal nerves.	Second to 12th dorsal segments.	In spinal diseases and progressive muscular atrophies.
Quadratus lumborum.	Deflects lower portion of spine laterally.	Lateral movements of lower vertebræ imperfect.	Lumbar nerves.		As above.
Abdominal muscles.	Aid in expiration; also in voiding bladder and straining at stool; in keeping vertebræ in position.	Lordosis, with protrusion of nates and abdomen; other actions deficient; cannot straighten up from recumbent position without assistance of hands.	Dorsal nerves.	Second to 12th dorsal segments.	As above.

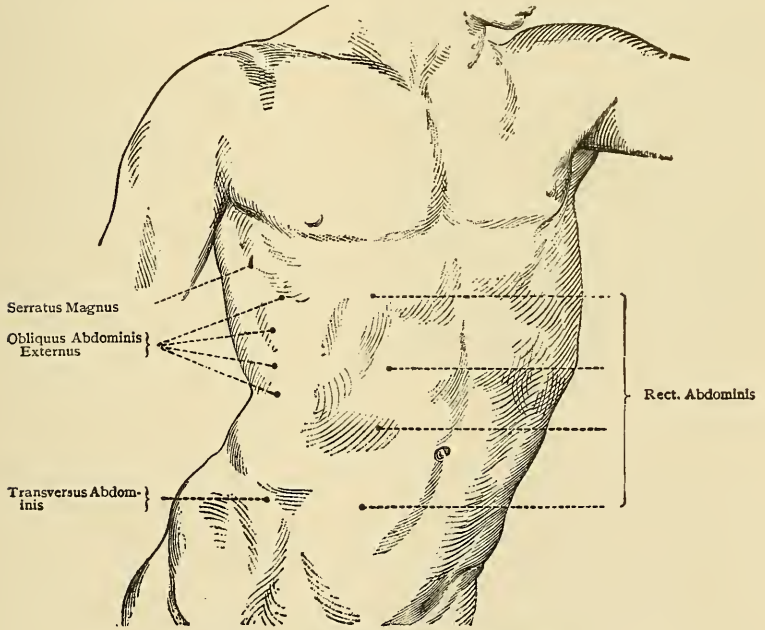


FIG. 29.—Motor Points of Trunk. (Bernhardt.)

Paralysis of individual muscles is not as frequent in the lower extremities as in the upper; large groups of muscles (the anterior tibial, the posterior tibial) are often paralyzed together. Symmetrical palsies are common (involving thigh groups, both post-tibial groups, etc.). Complete paraplegia of the lower extremities is seen in some spinal and cerebral lesions of children (spastic birth palsies, dorsal myelitis (poliomyelitis and myelitis), and in multiple neuritis. Paralysis may be flaccid (spinal or peripheral) or spastic (cerebral, or spinal). If spinal and spastic, lesion is in lateral column.

FIG. 30.—Patient showing Lordosis of Vertebral Column, due to Weakness of the Extensors (Erector) of the Spine.

To test the exact extent of paralysis, the child is to be placed on its back; if it does not move the limbs at all voluntarily, suspicion of very great paralysis or of excessive pain is a safe one.

If there is total paralysis, child cannot hold the leg, even if it be raised a few inches only; determine whether child can abduct, adduct, flex, or extend the various parts. Failure to abduct may be due to contracture of adductors; so also incomplete extension of knee may be due to contracture of flexors of knee. Contractured muscles must be considered with particular care. In very young children, if it is desirable to determine whether there is any actual paralysis, pinch the toe steadily until the child draws the leg away, or may be expected to do so.

With older children special tests can be made. Ask child to keep thigh flexed while you oppose; to test flexors of thigh, rest your hands on its knee and ask it to push it away by lifting up thigh; to test extension of thigh ask it to climb on a chair or observe whether it raises hip in walking. Place the child on a table and ask it to swing leg forward and backward; to keep leg straight while the physician endeavors to flex it (for extensors of leg), or to keep leg flexed while physician attempts to straighten leg (for flexors of leg). To test flexors or extensors of foot and toes, it should be asked to perform dorsal and plantar flexion with and without resistance.

To raise on tiptoes, or to raise toes while keeping heel on ground are good tests for the posterior and anterior tibial groups. Paralysis of certain groups is attended by contractures of others. Deformities of the feet (pes equinus, equino-varus, and valgus) result from paralysis and contractures of the flexors, extensors, and of the intrinsic muscles of the feet.

Examination of the gait is of the utmost importance; an attempt should be made to have the child walk; and even if it cannot walk, the position of the legs in standing, or in attempting to walk, is often quite characteristic. Cross-legged position and cross-legged progression occur in children (spastic paraplegia or diplegia); the gait may be paretic, spastic, ataxic, or a combined form of all three. If the gait is simply paretic the child drags its legs carefully, walking slowly and without raising the feet. The spastic gait is common in the hemiplegias and other cerebral palsies of childhood; also in spinal palsies (cervico-dorsal myelitis with involvement of the lateral columns). The knee is stiff, the leg is moved as a whole, and the child walks either on the ball of foot or on its toes. The ataxic gait, variously modified, occurs in neuritis in cerebellar disease, and in hereditary ataxy. In walking the child reels, its legs are moved extravagantly, and it walks with a broad base. A swinging gait is very frequent in poliomyelitis. The gait varies according to the muscles affected; in dystrophies, the gait is apt to be waddling, or a mixture of the waddling and swinging gait.

To be efficient, muscular action must be well directed. The contractions of muscles may be entirely normal, but unless the functions of muscles acting together are properly co-ordinated, the contractions may fail of their proper effect. In all movements (in walking, standing, writing, grasping, talking), co-ordination of the muscles therein concerned is essential. We speak of inco-ordination or of ataxia, if movements go wide of the mark. If a person, instead of touching the end of the nose with the tip of the fingers (eyes being closed), fumbles all over the face, he has ataxia of the upper extremities; and if instead of preserving his balance accurately in walking, a person sways from one side to the other, and to keep his balance at all walks with a broad base, that person has an ataxic gait. The ataxia may result not merely from inco-ordination of the muscles, but from insufficient sensory (muscular) impressions of the position in space of various parts of the body. If a person is asked to cross one knee over the other (with eyes closed), he must know exactly where the other knee is, if he is to perform the movement with nicety; and he must be able to gauge the exact extent of muscular action to

be employed. With the assistance of vision muscular co-ordination is more perfect. In children ataxia is less frequent than in adults; it occurs in multiple neuritis, in hereditary ataxy, and in cerebellar disease. (In the last-named, there is considerable reeling in addition to mere ataxia; there is inco-ordination and great uncertainty.)

Before leaving the muscular system it is well to note the tonus of the muscles. In children this is of importance; for by the mere handling of a leg the experienced physician will be able to determine whether muscles are normal, flabby, atrophied, or contractured. Every normal muscle, if struck lightly with a percussion hammer, or with a finger, will exhibit a wavelike contraction. This mechanical excitability may be increased, so that the entire muscle contracts forcibly upon the slightest tap (tapping the quadriceps femoris may produce movement like that of the knee-jerk); or instead of single contractions, small fibrillary tremors may be observed on tapping atrophied or degenerated muscles.

Sensory Disturbances are not easily determined in children. Many of the finer tests cannot be applied at all, for young children are not sufficiently intelligent to give reliable answers. The disturbances of sensation may be either subjective or objective.

1. Subjective sensations may vary according to character of sensory perception; thus we may have subjective sensations of heat, of cold, of pressure; if these sensations are intense, they may be equivalent to pain (Goldscheider); there may also be perverted subjective sensations, such as formications and other paræsthesiæ (feeling as though a part were swollen, enlarged, unusually heavy and the like). Pain may vary in character (boring, cutting, thumping, burning); its description, as Goldscheider has shown, depending very largely upon well-known sensory impressions that have been associated with pain. Distribution of pain will vary; it may follow the distribution of nerves (neuralgia and neuritis); it may be restricted to definite areas, and such areas of pain may hold definite relations to disease in distant parts (according to Head, Dana, Goldscheider, and others). Pain persistently referred to one spot is designated as Topoalgia.

2. In testing for objective disturbances of sensations, we must note that ordinary sensation is complex in character; that we have perception of touch, of pain, of heat and cold, of the active and passive movements, of pressure

and of resistance. There may be complete or partial loss of these various sense perceptions.

The simpler the tests the better; the æsthesiometer is superfluous. To determine tactile sense, passing cotton over the skin is generally quite sufficient; or still better, let the physician close his eyes and touch the patient ever so lightly; his own perception of contact will tell whether that of the patient is above or below his own; asking the patient to state in which direction a finger is being passed is another good test; a correct answer implies normal tactile perception at every point; writing numbers on the skin may be tried in older children. For pain, we may test by pricking with a pin, by using the faradic current, by pinching and the like. In all these tests the child's facial expression.should be studied carefully. To determine the temperature sense it is best to employ test-tubes with hot and cold water, or to place metallic objects on the skin and see whether differences in heat or cold can be made out. Goldscheider has shown that there are special areas in the skin for the perception of pressure, of heat and of cold; and that the acuteness of sensory perception varies in different areas.

His test for minimum or maximum heat and cold perceptions are not practicable, however, at the bedside—least of all in children.

The term "muscular sense" led to misconceptions. It included a perception of passive movements, of active movements; of pressure and of resistance, and a perception of the position of parts.

To test the perception of passive movements it is best to take the joint firmly between the two hands; then move the joint slightly and ask whether patient knows what has been done; a deficiency of this sense can be made out easily. This test is an important one, for many cases of ataxia are dependent upon the loss of this special perception. To test perception of weight, it is best to use rubber balls of same size, but filled with varying quantities of shot. The balls are placed on the hand, on the leg, etc., and the patient is asked to determine which is lighter or heavier. The patient's knowledge of the position of parts is tested by placing one part (a leg, a finger), in a definite position, or raising it to a definite height (eyes closed) and then asking patient to put a symmetrical part in relatively the same position.

Loss of sensation is termed anæsthesia; diminished sensation, hypæsthe-

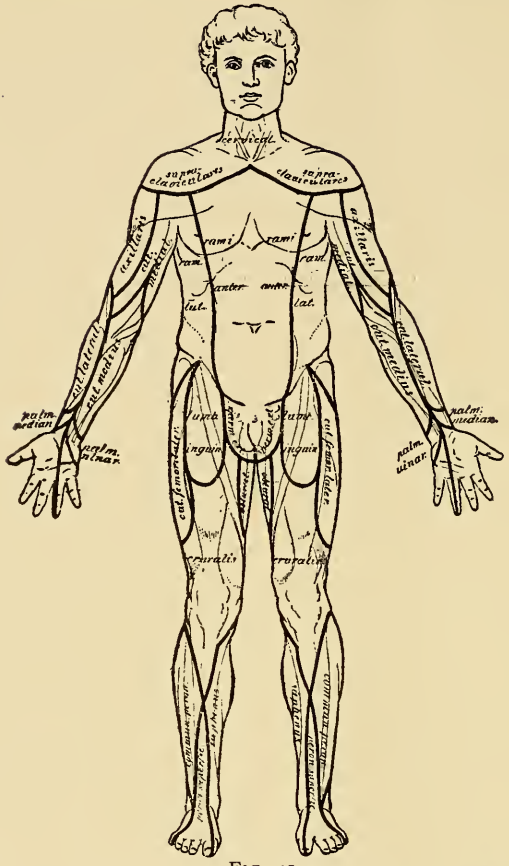


FIG. 31.

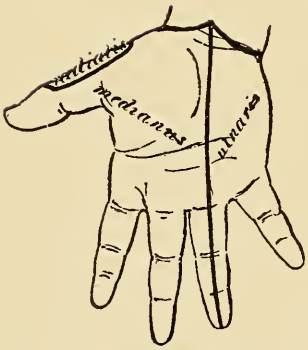


FIG. 32.

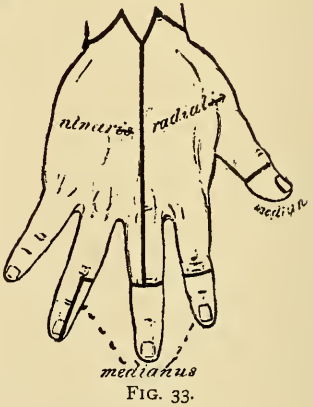


FIG. 33.

Distribution of the Sensory Nerves. (Freund.) Figs. 31–37.

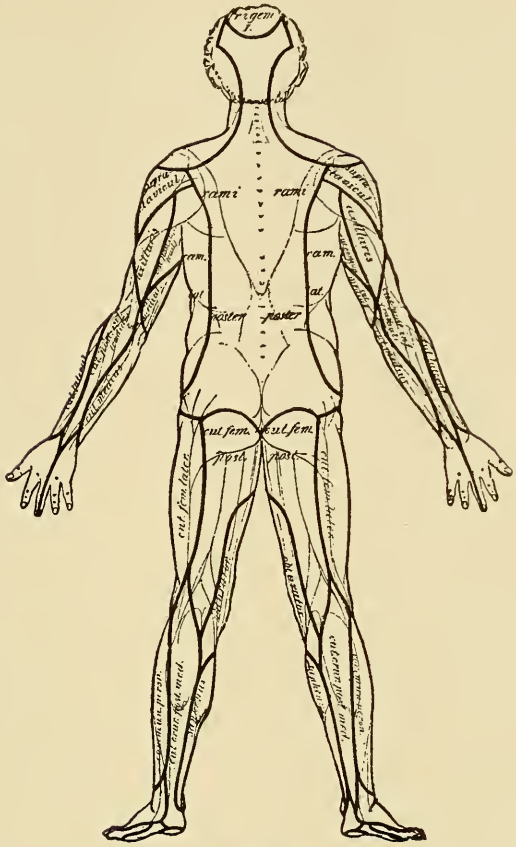


FIG. 34.

FIG. 35.

FIG. 36.

FIG. 37.

sia; and increased sensory perception is termed hyperæsthesia. Disturbances of sensation may vary with the distribution of sensory nerves (Figs. 6, 31–37). Hemianæsthesia implies loss of sensation in one half of the body. This is rare in children, and occurs chiefly in hysterical affections. Anomalies of temperature sense occur in some cord affections and in cases of neuritis. The stereognostic sense implies a correct perception and appreciation of an object through the cutaneous senses (with eyes closed the patient is to recognize familiar objects—such as coins, keys, buttons, placed on different parts of the body). Unless all sense perceptions and associated memories are normal, astereognosis may be developed.

Examination of Reflexes.—We distinguish between the deep or tendon reflexes, and the superficial or cutaneous reflexes. The former constitute the more important class. Among the deep reflexes the patella-tendon reflex or knee-jerk has the greatest clinical significance; next in order is the ankle clonus, or Achillis-tendon reflex, and in many cases the behavior of the wrist reflex, the triceps reflex, and even the jaw-jerk is carefully to be considered. In children it is more difficult than in adults to elicit the deep reflexes, for the former cannot relax their muscles easily, and thus inhibit to a degree every reflex movement. In testing for the knee-jerks and for other reflexes, it is best to take the child unawares, before it has learnt to know what the test means. While busying one's self with other parts of the body, or while talking to the child, if the leg is in a favorable position, strike the tendon smartly, and the reflex movement will ensue if the conditions are normal. If this first test does not succeed, seat the child at the edge of a table with its legs dangling, and then quickly strike the tendon. If the child continues to inhibit the reflex, I am in the habit of placing my left hand between the crossed knees of the child in such a way as lightly to grasp the posterior surface of the upper leg. The child is compelled to relax its muscles, and I can easily tell whether it makes the slightest effort to contract them. If the tendon is struck, the excursion of the leg is bound to follow, provided the reflex is present. In older children Jendrassik's method (clinching the fists while the test is made) may be employed; but it is still better for the physician to ask the child to squeeze his fingers the very instant he strikes the tendon.

To elicit the ankle clonus, it is best to perform sudden dorsal flexion of the foot, while keeping the knee resting on the other hand and in the position of moderate flexion. The same clonic movements can be elicited by striking the Achillis tendon while the foot is in moderate dorsal flexion. Babinski has suggested that the patient be asked to rest the leg on a chair; the Achillis tendon can then be struck with ease.

The wrist reflex is tested by striking the radial side of the forearm near the wrist; a smart tap will cause contraction of the supinator and biceps muscles. The triceps reflex is elicited by striking the tendon while the arm is flexed. The "jaw jerk," a reflex contraction of the masseter, is brought about by striking the middle of the chin when the mouth is slightly open, or by tapping a pencil laid on the lower teeth or jaw. I have seen this distinctly present in a case of multiple sclerosis in a child.

In testing the wrist or triceps reflexes, place the joints in a position of moderate flexion so as to give the muscles the fairest chance for contraction. The deep reflexes may be present, diminished or exaggerated. The knee-jerk is present under ordinary conditions; the mere presence of the ankle clonus is abnormal; and the reflexes in the upper extremity are present in some and absent in other normal individuals.

The deep reflexes are diminished or absent in peripheral nerve disease, in spinal-cord affections involving the posterior columns and the gray matter of the cord; in disease of the cerebellum. They are exaggerated in diseases of the brain and in diseases involving the lateral columns of the cord; also in some functional diseases.

The diminution of the reflex is determined easily enough; but the absence of the reflex is more significant than a mere diminution. The diminution or absence of a reflex may be apparent only. Thus the knee-jerk may be wanting in consequence of contraction of the posterior flexors of the leg, or there may be anchylosis of the joint preventing free movement; but in such cases the contraction of the quadriceps can be seen or felt upon tapping the tendon. In some children the tendon is displaced or so imbedded in fat that it cannot be struck readily.

It is more difficult to state when a reflex is exaggerated. In this matter comparison based upon experience is the best guide. We can safely speak of an exaggerated reflex if there is an exceeding quickness of the response, if the slightest tapping produces a liberal movement; or if clonic movement results from a single blow (as in patella clonus). If the reflex can be elicited indirectly by striking the finger placed over the tendon, the reflex is surely exaggerated. Moreover, if a child of three years or more is so seated that its feet are in solid contact with the floor, and if when its patella tendon is struck, the foot is lifted from the floor, the reflex is considerably increased.

Very lively knee-jerks do not necessarily imply organic disease, but exaggerated knee-jerks plus ankle clonus and a Babinski sign are indicative of this. Any change of the reflexes in one half of the body is always suspicious of

organic disease, and so is the association of increased mechanical excitability and of contractures with increase of the reflexes.

The superficial or cutaneous reflexes are not of great importance in childhood. The abdominal, epigastric, and cremasteric reflexes are but poorly developed in children; the last named is absent in many older children; and no significance can be attached to its presence or absence unless the condition is not the same in both halves of the body. The plantar reflex (retraction of the foot on irritation of the soles) deserves a few words of comment. Like the other cutaneous reflexes it is absent in diseases of the peripheral nerves, and in diseases of the spinal cord involving the course of the reflex arc; if the lesion is higher than the level through which the arc passes, the reflex may be exaggerated, but this is not as constant a feature as is the case with the tendon reflexes. In all unilateral brain-lesions the cutaneous reflexes are absent as a rule on the paralyzed side. The plantar reflex is generally absent in a condition of coma (as well as in deep sleep), and its presence or absence may give a clew as to the degree of coma in a given case.

The Babinski phenomenon (dorsal flexion of the big toe following irritation of the sole of the foot) is an important symptom pointing to involvement of the pyramidal tract; it is well, however, to bear in mind that in children under four years of age a dorsal flexion of the toes is the normal reflex.

ELECTRICAL EXAMINATION.*

The distribution of a paralysis, the flaccid or spastic condition of the muscles, the behavior of the reflexes, may enable us to make an accurate diagnosis in many cases, but in many more the diagnosis cannot be safely established until a careful electrical examination has been made.

The chief object of such an examination is to determine the response of various muscles and nerves to the faradic and galvanic currents, and to compare such response with the conditions obtaining during health.

The younger Westphal has shown that the nerves and

* The author cannot undertake to give a detailed account of medical electricity; for this the student is referred to the treatises of de Watteville, of Beard and Rockwell, of Erb, Lewandowski or Toby Cohn; also to the special chapters in the text-books of Gray, Dana, and Gowers. The importance of a thorough knowledge of electricity should not be overlooked. Whatever one may think of the value of electricity as a therapeutic agent, there can be no question of its great merit as an aid to diagnosis. Moebius argues that the experienced neurologist can make a diagnosis without the assistance of electrical tests, and that such tests are not wholly reliable. But they are reliable in fully ninety-five per cent. of all cases, and as for making a diagnosis without the aid of electricity, that may be possible. A clever neurologist can also diagnosticate a cerebral tumor often enough without the ophthalmoscope, yet he will not spurn the corroborative evidence which an examination of the papillæ may furnish.

muscles of the new-born, and of children up to the age of five weeks, do not respond except to very strong currents, and even then the contractions are slow. But after that age the normal nerve and muscle will respond to faradic and galvanic stimulation. A knowledge of the position of the motor points is essential. (See Figs. 7, 13, 14, 15, 19, 20, 28, 29.)

Always begin the examination with the faradic current, and apply the one pole to the nerve or muscle to be examined, and place the other pole at a safe distance, say on the sternum. Use mild currents (children grow very restless under strong currents), and determine the weakest current with which a contraction is to be obtained, or whether any contraction can be obtained at all, or not. Place the arm or leg in a position favorable for contraction of the muscles to be tested, and make sure that the contraction which ensues is the one that should naturally follow. Do not, for instance, claim that the electrical conditions are normal if on applying an electrode to the extensor digitorum communis, a flexion contraction of wrist and fingers ensues. And if there is any doubt as to the strength of the current, let the physician apply it to himself; he will be more certain to be merciful if he does; or if there is a doubt whether a nerve or muscle should respond to a given strength of current, let that same current be applied to the same point in the opposite half of the body. Thus we can compare two peronei with each other; or we can compare a peroneal nerve with a facial or a median nerve of the same or opposite side of the body,* or we can compare the degree of excitability with the figures given in tabular form by Stintzing for the minimum and maximum currents required to produce contractions of various nerves and muscles.†

A simple diminution of faradic and galvanic response of nerves and muscle occurs in light cases of peripheral neuritis, in hysterical conditions, in atrophies following joint disease or from disuse, and in some primary dystrophies. A simple and continued increase of response is observed practically only in cases of tetany. But in addition to mere quantitative changes there are also changes in the character and quality of the response to be observed in cases of degenerative disease of the peripheral nerves, of the muscles, and of parts of the spinal cord. The changes implying degenerative disease are referred to as the reaction of degeneration.

* In children the motor points may be obscured by deposits of fat.

† American faradic batteries are now provided with a sliding scale which enables the physician to record how far apart the primary and secondary coils were when first contraction was obtained. In recording the effect of the galvanic current, the strength should be stated in milliampères.

The following table will give the salient features of normal electrical conditions and of the reaction of degeneration:

Normal Electrical Conditions.		Reaction of Degeneration.	
	Nerve and Muscle.	Nerve.	Muscle.
Faradic current.	Contractions good; prompt.	No response (except in partial R. D.).	No response (except in partial R. D.).
Galvanic current.	Contractions prompt and quick.	No response.	Increased excitablity at first, then diminished; contractions sluggish.
Order of contractions.	1. K. C. C.* 2. A. C. C. } interchangeable. 3. A. O. C. } 4. K. O. C. 5. K. C. Te. (rare).		With stronger currents. 1. A. C. C. } A.C.C.>K.C.C. 2. K. C. C. } 3. A. O. C. 4. K. O. C. or A. C. C. = K. C. C.

* K. C. C. = Kathodal closure contraction; A. C. C. = Anodal closure contraction; A. O. C. = anodal opening contraction; K. O. C. = Kathodal opening contraction; K. C. Te. = Kathodal closure Tetanus.

The preceding table states that in the reaction of degeneration (R. D.) the nerves and muscles fail to respond to the faradic current, but that the muscles continue to respond to the galvanic current in altered fashion; first of all, the contractions are sluggish, and this is the most important point, and secondly, the order of contractions is reversed; the anodal closure contraction can be obtained with weaker currents than are required for the kathodal closure contraction.

The mistake is commonly made to suppose that there is no reaction of degeneration present unless the galvanic order of contractions is reversed; this occurs in the majority of typical cases, to be sure; but the failure of response to the faradic current is early evidence of a reaction of degeneration, and often precedes the development of abnormal galvanic conditions.

In first examinations the faradic test is therefore much the more important.

Many recent authors (including Remak) agree furthermore in considering the sluggishness of contraction much more significant of degenerative changes than the reversal of the normal formula.

It should be remembered also that muscles may be so much atrophied as to give no contraction to the strongest galvanic current.

A partial reaction of degeneration is often overlooked; it implies a mere diminution of faradic excitability of nerve and muscles, but the response of the muscle on direct galvanic excitation is slow, and the formula may be reversed. Even if the faradic response is present, it is important to make sure that the galvanic response is prompt, not sluggish.

The greatest service which electricity affords us in diagnosis is in differentiating between cerebral diseases on the one hand and certain spinal and peripheral diseases on the other. The ganglion cell of the anterior horn and its analogue in the cranial nerve nuclei are responsible for the normal electrical condition of peripheral nerves and muscles. Lesions involving this cell, or interfering with the transmission of impulses from it to the periphery are attended by changes in electrical reactions; in all other diseases the electrical conditions are practically unaltered.

Whence it follows that the electrical conditions are:

Normal in	Distinctly Altered (R. D.) in
1. All cerebral diseases,* excepting those of cranial nerve nuclei.	1. Bulbar paralysis (acute and chronic).
2. Diseases of lateral and posterior columns of spinal cord.	2. Polioencephalitis superior (Wernicke).
3. Functional troubles.	3. Poliomyelitis (acute and chronic).
4. Mild peripheral troubles.	4. Progressive amyotrophies.
5. In some forms of muscular dystrophy.	5. Amyotrophic lateral sclerosis.
	6. Myelitis, but only in muscles represented in diseased level.
	7. Gliosis and tumor involving gray matter.
	8. Anterior root disease (syphilis, tumor, etc.)
	9. Vertebral disease (Pott's disease, tumor).
	10. Peripheral neuritis (traumatic, rheumatic, toxic).
	11. In some forms of muscular dystrophy.

* A very few exceptions have been reported.

To complete the examination, be sure to look for trophic symptoms which occur in many functional disorders as well as in disease of the peripheral nerves and in some diseases of the spinal cord (gliosis, tumor); and finally the conditions of the vesical and rectal reflexes should be

recorded; though in regard to them the influence or lack of previous training of the child must be taken into account. A sudden loss of control is often associated with serious cerebral and spinal disease.

Lumbar Puncture is a recent addition to our methods of examination. It is practised for the purpose of withdrawing some of the cerebro-spinal fluid. The examination of this fluid may render valuable aid in the diagnosis of cerebral and cerebro-spinal affections. Its value as a therapeutic measure is still in doubt.

Lumbar puncture was introduced by Quincke. His directions are as follows: The patient is to lie on his left side, near the edge of the bed, with knees well drawn up. The trocar is to be inserted in the median line between the third and fourth or between the fourth and fifth lumbar vertebræ (Fig. 38). After reaching the dural sac the trocar

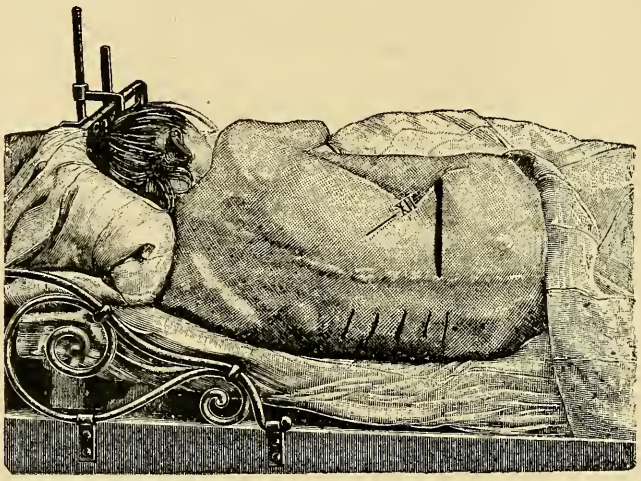


Fig. 38. (After Quincke.)

Slightly Modified; the Horizontal Lines Correspond to the Lumbar Vertebræ.

needle is withdrawn and the cerebro-spinal fluid will flow out. Quincke advises connecting the canula with a small rubber tube and a glass manometer. If there is no object in measuring the pressure at the time of tapping, the trocar is sufficient and the fluid may be received in a test-tube.

To determine the site for the puncture, draw a line at a tangent to the crest of the ilium; this will strike the fourth lumbar space.

General or local anæsthesia is unnecessary except in unruly patients. Do not withdraw more than 20 to 30 c. c. at a sitting. Use sterilized test-tubes.

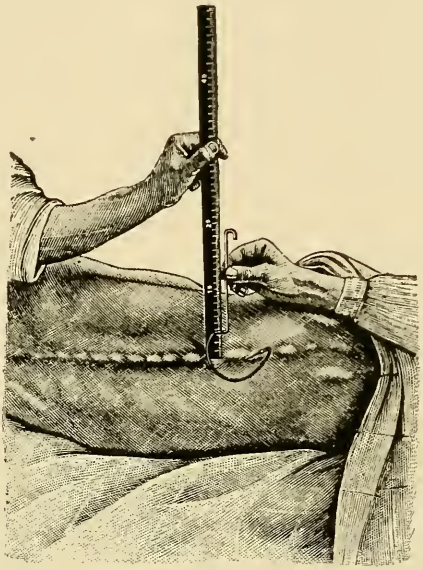


FIG. 39. (After Quincke.)

The procedure is a harmless one except in cases of cerebellar tumor and in neoplasms in the vicinity of the fourth ventricle.

PART I.

GENERAL NERVOUS DISEASES.

CHAPTER II.

CONVULSIONS—ECLAMPSIA INFANTUM.

THE peculiar seizures so common in infancy, and designated as convulsions, constitute a symptom, not a form, of disease. The unusual frequency of convulsions in early childhood points to the greater excitability in the child of the motor mechanism of the brain. The motor centres in the cortex are more apt to "discharge," and the inhibitory power of the brain is less developed than in the adult. Eclampsia has been found to be a convenient term for the condition in which convulsions are apt to occur, but it should be remembered that it is neither a disease nor a sufficient diagnosis in any given case, for eclampsia in children, as well as in adults, may be the result of many widely different disorders.

However frequent these convulsive seizures may be in early life, healthy children are as exempt from them as are healthy adults. If they are not the result of organic disease of the brain, their occurrence indicates functional involvement of the brain in connection with disease in other parts of the body.

A convulsion is a motor discharge resulting in muscular contractions of one or more parts of the body. In the majority of cases these convulsive movements are associated with a number of other symptoms; viz., an initial cry, turning of the eyes upward or inward, very sudden loss of consciousness, involuntary passage of urine and of fæces, prolonged drowsiness, and a condition of stupor or of coma. The muscular movements may be tonic at first, but are apt to lead to clonic spasms. No two convulsive seizures are exactly alike, but the student who has seen a single seizure will not forget the main features of the condition. The

convulsion may come on without warning, or the child may have complained of uncomfortable sensations in the head or stomach, of a little dizziness or of nausea; the child gives a shrill cry and at once is thrown into tonic and clonic convulsions. These may be partial or general, unilateral or bilateral. The legs are stiff, the arms bend at the elbow, the wrists are flexed, and the hands clenched, with the fingers firmly closed upon the thumb. The head is thrown back or rotated to one side; the back may be arched, the face is pale at first, but soon reddens and the eyes remain wide open. The pupils do not react. In older children the tongue may be caught between the teeth, and bloody

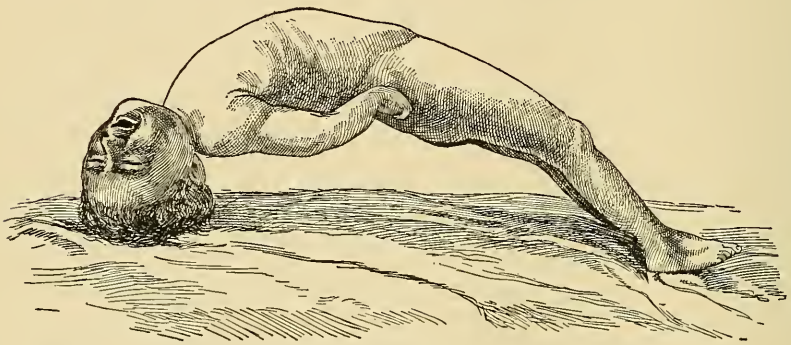


FIG. 40.—Child of One Year Photographed During a Severe Convulsive Seizure.

froth may be noticeable at the lips. In very young children we have the ordinary foam at the mouth. The respiratory muscles are in a condition of spasm; the child often turns blue rapidly and is in danger of asphyxia. Fortunately the tonic spasm soon ceases, slighter clonic convulsions then set in, and after a little the child grows quiet, and is certain to lie in a dazed or stuporous condition for a period that may vary from several minutes to as many hours or days. While the clonic spasm lasts there is imminent danger of injury to the child's head and limbs in consequence of the severe jactations, but this danger is not as serious as that from asphyxia during the period of tonic spasm.

CAUSES.—Convulsions always denote "cerebral (cortical) irritation." It might be sufficient to state that any morbid

process which brings about cortical irritation, directly or indirectly, is liable to cause convulsions. But it will be better to enumerate the chief conditions under which infantile convulsions occur.

I. Convulsions occurring within the first few days of life are, as a rule, the result of meningeal hemorrhage, due either to protracted labor, or to instrumental delivery. If the child survives, the injury done to the brain often leads to the development of spastic palsies. (Cf. chapter on Cerebral Palsies.)

II. The convulsions may be due to organic disease of the brain, such as tumor, abscess, meningitis, or vascular lesions; in all of these cases they may be the earliest symptom of the disease, but other symptoms associated with them, such as headache, paralysis, optic neuritis, and the like, will be forthcoming.

III. They may mark the onset of any acute infectious disease. The initial convulsion in children may have the same value as the initial rigor in the adult. The former, with or without fever, is the clinical equivalent of the latter. It is common at the onset of pneumonia, scarlet fever, and measles; and I have also witnessed convulsions in malarial fevers, and at the onset of chicken-pox. The general practitioner is very apt to err in the interpretation of such convulsions. A child that has had a convulsion, preceded or followed by a rise of temperature, is often suspected to be in the first stages of a meningitis or of some other cerebral disease. While this is true in some instances, the fact is entirely overlooked that the seizure may be the initial symptom not only of any of the ordinary infectious diseases, but of the acute spinal diseases as well—poliomyelitis, for instance (probably an infectious disease).

IV. The convulsions may be of reflex origin. Almost every possible peripheral disturbance has been supposed by one author or the other to be a sufficient cause of convulsions. I need merely mention ocular insufficiencies, nasal obstruction, narrow prepuces, not to call up a dozen or more about which there has been much, and often bitter, discussion. Of the influence of two conditions there can be no reasonable doubt. The first of these is dentition; the

second, gastro-intestinal irritation. There has been some question whether a tooth about to cut through the gum can cause an eclamptic seizure. Delayed dentition is so frequently associated with rickets that the latter condition is considered by many to be the chief factor, and this I believe to be true; but convulsions do at times occur in children who present no tangible signs of rickets. Moreover, there is good reason why a cutting tooth should produce convulsions in a child so disposed, say by rickets, or any form of exhausting disease. "The cutting tooth" is a direct irritant to the filaments of the trigeminal nerve, which carries this irritation easily enough to the convulsive centres at the base of the brain. I have little doubt of this special reflex origin of convulsive seizures since observing in the adult the occurrence of severe epileptic paroxysms in cases of trigeminal neuralgia.

The influence of gastro-intestinal irritation is well illustrated by the convulsions occurring in the course of an acute or chronic intestinal catarrh, in the ordinary summer diarrhœa of young children, or with the exhausting chronic diarrhœa in older children. The same phenomena are apt to occur in the presence of animal parasites (pin-worms, and, above all, tape-worms), and I have known severe convulsions to disappear promptly after the removal of a tape-worm in children between the ages of four and eight years never to return again.

V. Convulsions may be due to poisons (organic or metallic) circulating in the blood. Under this heading we may include the convulsions of uræmia, those due to the presence of ptomaïnes in the stomach and bowels, and convulsions produced by the administration of lead and other metallic poisons. Toxæmic convulsions are not as common in the child as in the adult, unless, following recent studies by Chenbach and others, we consider the convulsions of infectious fevers and of intestinal troubles to be due to some form of auto-intoxication.

VI. Convulsions may result from severe loss of blood, from any exhausting disease, or from such constitutional disturbances as scrofula and rickets. A vast literature has grown up on the subject of rickets and convulsions. What-

ever theory one may be willing to adopt in order to explain this close relationship, the fact remains that an enormous percentage of children having convulsions are affected with rickets.

An hyperæmic condition of the cortex analogous to the condition of the bones in rickets, is the only explanation we can give of the frequency of convulsions during rickets, unless we are satisfied to accept the very vague statement that the latter is a simple predisposing cause of convulsions.

VII. A convulsion may be idiopathic, hereditary, or, if you choose, the first incident in the course of an ordinary epilepsy. This possibility must be kept in mind, but it is a more probable explanation if the child happens to be above, rather than under, four or six years of age. This interpretation of a convulsive seizure occurring in a child should be given only after excluding every other possible cause.

It is surprising, but a fact nevertheless, to which I have called attention a number of times, that the more carefully we scrutinize cases of convulsions, or of epilepsy for that matter, the fewer of them appear to be truly idiopathic. It will be well not to make the diagnosis of an incipient epilepsy unless we are informed that the child has had previous convulsions at somewhat long intervals, or unless we have occasion to observe subsequent convulsions without a distinct additional cause for each seizure.

VIII. Convulsions may be due to traumatism. In some cases actual hemorrhage over the motor centres is the immediate cause of the convulsions. In other cases no tangible injury to the brain has resulted, and we are therefore compelled to regard the spasm as the expression of shock.

THEORIES.—Many theories have been advanced to explain the occurrence of convulsions. No theory is entirely satisfactory; but by physiological experiments and the experiment of disease we have learned to know the conditions under which convulsions are most apt to occur.

There are, first of all, the time-honored experiments of Kussmaul and Tenner, showing that convulsions occur in a rabbit after suddenly tying the cervical arteries and thus cutting off the blood-supply. Their conclusions are op-

posed to the older theories that these seizures were due to an hyperæmic condition of the brain. The question arises whether the sudden withdrawal of blood deprives parts of the brain of their functions, or whether it simply acts as an irritant to the cortical centres. The latter explanation seems plausible, in view of the experiments of Hitzig, Ferrier, Horsley and others. A mechanical irritation of the pons (floor of the fourth ventricle), as proved by Nothnagel, also produces convulsions. It is evident that toxic agents have the same effect as the electrical or mechanical irritation just referred to, else we could not explain the convulsions due to toxic agents, those of uræmia, for instance, and those due to asphyxia from the accumulation of carbonic acid in the blood.

Every morbid process in the motor areas of the cortex, if not absolutely destructive, is apt to cause convulsions; but what is the relation between the convulsive centres in the cortex and Nothnagel's epileptic centre in the pons? Does the cortex contain an actual convulsive centre? If you irritate the lower epileptic centre in an animal whose hemispheres have been divided from the rest of the brain you will get convulsive spasms of the entire body. The lower centres have the power, therefore, of "starting" convulsive seizures, and it is probable that they are under higher control in the fully developed brain, and so long as normal conditions exist. It is this power to control, this inhibitory force, that resides in higher centres. The removal of this inhibition through disease of such centres liberates the energy of the lower centres.*

Age and Frequency.—By far the largest majority of convulsions occur during the first two years of life. The Philadelphia Health Reports, as condensed by Lewis, give ample proof of this.

Seven thousand five hundred and eight deaths among

* Neurologists, the world over, have adopted Hughlings Jackson's views without endorsing every detail of his theories. Jackson considers the central nervous system to be made up of three tiers of sensory-motor centres. All parts of the body are represented in each tier, but the higher centre controls one or both below it, and has the power to inhibit the discharges of a lower centre. In a young child the higher tiers do not exert a controlling influence, since they are imperfectly developed; hence the discharges are so much more frequent than in later years.

minors (during period of 1876–1883 inclusive) were due to convulsions and laryngismus stridulus. Of these were:

	Under 1 year.	1 to 2 years.	2 to 5 years.	5 to 10 years.	10 to 15 years.	15 to 20 years.
Cases of convulsions....	4,993	1,335	893	178	38	26
Cases of laryngismus stridulus.....	24	7	9	5	..	..
Total	5,017	1,342	902	183	38	26

While the statistics illustrate a general truth, some comment is necessary. First, convulsions occurring at this early age are not necessarily fatal, although those occurring in the first few days after birth are much oftener fatal than those occurring later. The brain has less power of resistance, and the processes giving rise to these early convulsions are a more serious menace to the life of the child.

Secondly. It should not be forgotten that convulsions are so much more frequent during the first two years of life because the diseases and conditions causing convulsions are most frequent at this period.

We must take into account an injury inflicted upon the brain during labor, defective brain development, dentition, the acute infectious diseases, and almost all the infantile cerebral palsies and gastro-intestinal disorders.

Convulsions may, however, occur at any age; but since the child's brain grows less irritable the older it grows, a convulsion occurring later in life often signifies more serious trouble.

The frequency of convulsive attacks in any given case will also depend upon the nature of the disease, of which it is merely a symptom. In the case of convulsions due to dentition, a convulsion or a series of convulsions may occur with the cutting of each tooth; if due to gastro-intestinal irritation, convulsions may recur until the condition is improved or relieved. In acute infectious diseases we are apt to have but one initial convulsion, or at most two or three distinct attacks; if there were more than a few convulsions during an attack of an acute infectious disease, I should suspect some cerebral complication.

It is a fact, important and well worth remembering, that

the initial convulsion is as little apt to be repeated as is the initial rigor. The first toxic invasion, and not the fever, seems to cause the convulsions. I have often observed convulsions with relatively low temperatures (103° F.) at the onset, without any repetition of the same during the course of an acute disease, even though the temperature reach 105° F. and over. I do not mean to deny, however, that high fever alone may cause convulsions, but it is not a common cause.

I have often had occasion to remark that *repeated* convulsions are characteristic of cortical disease, and this is borne out by the cerebral diseases of childhood.

In the majority of such cases the convulsions will be but one of a series of symptoms; and if convulsions occur repeatedly without any further evidences of cerebral disease, the suspicion of true epilepsy must be entertained.

Symptoms.—It may seem odd that we should treat of the symptomatology of convulsions, which we have termed a symptom and not a disease, but the sequence of events during a convulsion is subject to great variations, and on this head there is need of further remark. A convulsion may be partial or general. Partial convulsions in the majority of the cases denote organic disease of the brain, and in this sense may be more serious than general convulsions; a slight convulsive twitching of a thumb may seem a small matter indeed, but it may be the first sign of a cerebral tumor which is bound to be fatal before long; while, as regards danger to life, a severe general convulsion may be entirely harmless. Partial convulsions are often unilateral, and sometimes limited to a single member.

The cortical centre of the part convulsed is the chief site of the lesion. Partial convulsions often become general; it is of importance, therefore, to inquire whether convulsions that affect all parts of the body start in any one particular member, or whether the person who is subject to general convulsions ever had unilateral partial convulsions.

All the phenomena of a convulsive seizure need not to be exhibited in each attack. We may have convulsive twitchings without loss of consciousness, and loss of consciousness without twitching movements. I have clearly in mind a child about three months old, whom I had reason to see

some nine years ago. Without any known cause the child was in a "faint spell," during which time the skin was pale, and the pulse became slow and irregular. Such a spell occurred about the same hour on successive afternoons, and later on the child had several each day. No other symptom could be made out, with the exception of a distinct enlargement of the spleen. The attacks were evidently malarial and yielded quickly to large doses of quinine. The child has had no convulsions since.

There is some difference of opinion among authors as to whether a convulsive attack begins with clonic or tonic movements, and which preponderate. It is well to know that, according to the researches of Unverricht and others, tonic and clonic convulsions are different in degree and not in kind—a tonic convulsion being practically a rapid succession of clonic movements. In children tonic spasm is supposed to precede clonic spasm, and in this respect to differ from the epileptic paroxysm of later life; but the distinction is of no importance, and, moreover, I have seen many spasms in children which were purely clonic from beginning to end.

One special form of convulsive seizure in children deserves special mention. This is

Laryngismus Stridulus.—Cerebral croup, inward convulsions, child-crowing, etc. The many synonyms are indicative of the frequency of this condition, which occurs more often in European countries than here, owing probably to the greater spread of severe forms of rickets in England and on the Continent.

In its simpler form laryngismus stridulus implies merely a spasm of the glottis. A child that may have exhibited an occasional crowing, croaking noise, is seized quite suddenly with severe spasm of the adductor muscles; the body becomes rigid, the head is drawn back, the face grows pale and then livid. In a few seconds the spasm relaxes, a deep inspiration follows with a hissing sound, and all is over for the time being, except that the child presents a haggard, tired look. Several such spasms may follow quickly upon one another. After the attack is over the child often vomits, is badly frightened, and from sheer fatigue falls

into a more or less natural sleep. It is not rare to have as many as twenty and more of such attacks during the course of twenty-four hours; in other cases single attacks follow each other at much longer intervals. It is quite exceptional, however, to have but a single spasmodic attack.

Other convulsive actions are apt to be associated with the laryngeal spasm. The diaphragm and other respiratory muscles are often convulsed; the fingers are firmly clenched, and the toes are in extreme plantar flexion ("carpopedal" spasm); general convulsive movements, and a total loss of consciousness may be developed before the seizure is over, proving the close relationship between the laryngeal spasm and general convulsions.*

CAUSATION.—There was much dispute among the older authors as to the true cause of laryngeal spasm. An enlarged thymus gland pressing upon branches of the vagus was supposed to be the most frequent cause; but this has been entirely disproven by the autopsies of Henoch and others, who showed that there was no correspondence between the state of the thymus and the occurrence of "internal convulsions." An overpowering weight of opinion is in favor of a close causal relationship between rickets and laryngismus stridulus. The researches of Elsässer, of West, Gee, Gay, Henoch, Jacobi, Heubner, and many others have offered substantial proof of this position. Of late years there has been a disposition to discredit this theory. Loos has denied the influence of rickets, while endeavoring to establish a relation between this convulsive disorder and tetany. To my mind the only proper relationship between the two is that they are both due, safely enough, to the influence of rickets. Loos has been severely criticised, and his facts have been disproven by Kassowitz, whose large experience of rickets has stood him in good stead.

Rachitic softening of the skull (craniotabes) has been supposed by Jacobi and others to be the active cause of laryngeal spasm. That the association of these conditions is unusually frequent cannot be doubted. Kassowitz states that only 48 of 370 cases of laryngeal spasm did not present

* Children suffering from laryngismus stridulus are prone to the ordinary eclamptic fits, thus furnishing further evidence of the close relationship between the two conditions.

marked craniotabes; but of these 48 cases 47 presented some other decided symptom of rickets. The fontanelles were closed in only 4 of these 370 cases, although 110 of these children were over one year of age. When rickets is not present the fontanelle should be closed at the end of the first year (Kassowitz).

But is a softened skull the direct cause of laryngeal spasm? Scarcely. The centres for vocal movements are far away from that part of the brain which is apt to be compressed in cases of craniotabes. The hyperæmic condition of the brain in rickets is of a piece with the hyperæmic condition of other structures and it is this hyperæmia which causes an unusual irritability of the centres which would not under ordinary circumstances "discharge" upon the slightest peripheral irritation (cold air, dentition, slight gastro-disturbances). Indeed there may be spontaneous discharge of these centres without peripheral irritation.

Laryngeal spasms occur most frequently between the sixth and eighteenth months, the very period during which rickets is most marked, if present, though it occurs often enough up to the age of three years and later. Patients subject to attacks are most apt to suffer from them during the coldest months of the year, viz., January, February, and March.

Much has been made of the supposed relation between laryngismus stridulus and tetany. Cheadle believes the two to be identical. Facial contractility may be present in cases of laryngismus, but cases of laryngismus in which the Trousseau symptom (spasm induced by compression of the artery in a limb) can be elicited during the interval between attacks are surely rare.

There is another form of convulsive seizure in children which reminds one of laryngismus. Many years ago a child, aged eighteen months, apparently healthy, was brought to me that would hold its breath in many a crying spell; after holding its breath for a few seconds it would turn blue, and its head would drop forward as it lost consciousness. After a few more seconds the child recovered consciousness, and all was well again until the next crying spell, when the same sequence of symptoms would be apt to occur. Treat-

ment by bromides and small doses of chloral put an end to these attacks in a few weeks.

Diagnosis of Convulsions. — It is scarcely credible that any one who has ever seen a convulsive seizure can mistake it for anything else. And yet it has happened within the author's experience that he was called to a case of supposed tetany, which was, however, nothing more than an ordinary convulsive attack.

The difficulties of diagnosis that arise are concerned altogether with the possible etiological factor in a given case. The physician who witnesses a convulsive seizure will do well to be reserved in giving an explanation of the origin of such seizure until he has carefully examined into the previous history of the case; and if he does this he will be able to say with great certainty whether the convulsion is due to some reflex irritation, to some cerebral or spinal disease, or whether it is the initial symptom of an acute infectious disorder. It is of the greatest practical importance, however, to be able to say distinctly whether the convulsion is the expression of functional disease or of organic disease of the brain. General convulsions are, in the vast majority of cases, of functional origin. Partial convulsions are, as a rule, the result of organic disease; but a child that has general convulsions may at some previous stage of its history have exhibited partial seizures, or a general convulsion may have begun in a strictly localized fashion. The part first convulsed, or the part alone convulsed, indicates that the centre in the brain which governs the movements of this special part of the body is the chief site of the lesion. A twitching movement of the thumb, so slight that many might be tempted to regard it as a trifling nervousness, indicates disease in or near the centre governing the movement of the thumb. A convulsive seizure, beginning with a twitching of the eyelid, with the drawing up of one corner of the mouth, is certain to be the result of disease in the respective centres of these parts.

The gravity of a convulsive seizure will depend very much upon the cause of the attack. In general terms a partial epilepsy pointing to organic lesions of the brain is a more serious matter than a general epilepsy, which may be func-

tional; but if a general convulsion is the expression of a typical epilepsy about to be developed, its importance is as grave as though it were due to gross disease of the brain. Convulsions occurring at the onset of acute diseases are much more apt to pass off without leaving a trace behind them than are those convulsions which occur during the further course of the disease.

There is considerable difference of opinion among authors regarding the remote dangers of convulsions. Many are inclined to believe that convulsions are always symptomatic, and that they rarely constitute a serious danger to the child's life; but others, like myself, relying chiefly upon a careful study of infantile cerebral palsies and of epileptic convulsions in the adult, are confident that convulsions are capable of, and often do give rise to, serious disturbance. This is not surprising if we consider that extreme cyanosis, with an accompanying intense hyperæmic condition of the brain, occurs during the acme of the convulsion. I have myself recorded a case of a child dying in an epileptic convulsion, in which at the post-mortem table a widespread recent subpial hemorrhage over the convexity and at the base was found to have been the immediate cause of death. Eustace Smith refers to a case with hemorrhage over the base, evidently the effect of convulsions, while Money and others have reported cases in which a palsy, setting in after a convulsion, was evidently due to hemorrhage from the smaller vessels of the brain coming on during, and caused by, a convulsive seizure. The possibility of such occurrences makes a convulsion a serious matter. It must be the earnest endeavor of the physician in attendance on the child to bring a convulsive seizure to an end as soon as possible. The longer a seizure lasts, the more frequently it is repeated, the greater the danger becomes—if not to the life of the child, at least to its future mental condition.

Hence it follows that we must be guarded in giving a prognosis of the child's condition after the seizure is over, the majority of them leaving no unfavorable trace upon the child, but every now and then cases occur in which the convulsions were the first of a long series of disasters. I shall never forget the case of a bright child who had a con-

vulsive seizure at the onset of chicken-pox; a rapidly developing idiocy was the result. Seizures occurring in the first few days after birth, inasmuch as they are the expression of serious disease in the brain, give a particularly unfavorable prognosis.

TREATMENT.—The first object of treatment should be to check the convulsion. Many remedies will have been applied long before the physician has arrived at the bedside of the patient. Hot baths, mustard poultices over the spine, the abdomen, to the soles of the feet are still in vogue and urged not only by knowing mothers, but also by many physicians (by Meigs and Pepper, for instance); but all these measures are to be deprecated, for they imply a loss of valuable time. If a convulsion ends while the child is in a hot bath it does so because it has run its course.

To check the convulsion inhalations of chloroform, or of chloroform and nitrite of amyl in equal parts, are much to be preferred to other forms of medication. A few drops poured on a handkerchief and held to the nose of the child are generally sufficient to diminish the severity of the convulsive seizure, and the nurse or physician sitting by can keep up this method of administering the drug until the tendency to convulsions has for the time being entirely disappeared. Recognizing the seriousness of convulsions, the physician will do well to wait until all danger from further attacks has passed. After the acute stage is over small doses of morphia (gr. $\frac{1}{10}$ to $\frac{1}{6}$) or of opium, according to the age of the child, may be given; and after the first day has been passed safely I should advise the discontinuance of the opiates and the substitution of small doses of bromide (3 to 5 grs.) several times a day, or small quantities of chloral hydrate (gr. v. to x.), either by mouth or in the form of a rectal enema.

Absolute quiet is the first essential of treatment. If there is good reason to believe that the convulsive seizure was due to gastro-intestinal irritation, or to the presence of intestinal parasites, steps should be taken to purge the bowels thoroughly, say by the use of castor-oil, calomel, and the like, or by appropriate treatment for the removal of worms. As soon as the cause has been removed, treat-

ment by small doses of opium, bromide, or chloral, will be in order for at least a week or ten days after the occurrence of the seizure.

If the convulsive attack is associated with fever, there is of course every reason for giving a tepid bath, with subsequent cool sponging. The reduction of the temperature will, to a certain extent, diminish the probability of repeated attacks. Should a child be seized with convulsions while in the bath, it should be removed at once and made to inhale chloroform and nitrite of amyl. It is far better in the cases of gastric disturbance to evacuate the stomach through the bowels than to attempt to give emetics, which of course cannot be readily swallowed, or to give hypodermic injections of morphia as suggested by Steffen. For to cause a child to vomit that has a tendency to convulsions is to increase the danger to which it is exposed.

Cases of laryngismus stridulus can be treated on very much the same principle as convulsions, excepting that the child should not be allowed to remain in the recumbent position; it should be carried about, and if breathing is interfered with, forcible means, such as pulling forward of the tongue, slapping of the chest with cold and wet towels, should be resorted to in order to bring about regular, rhythmical breathing.

If the convulsions are due to rickets, very active treatment, looking toward the improved nutrition of the child, will be necessary. It should be placed upon the albuminoids, and only small quantities of the cereals allowed. Moreover, in these cases, Kassowitz's plan of administering phosphorus with cod-liver oil, as in the scrofulous diathesis, is of greatest importance. Syrup of the iodide of iron, or any of the many preparations of iron now to be obtained in the market, can be administered. If the attack is the beginning of an epilepsy, no time should be lost in placing the child upon the regular treatment of this condition, for the details of which the student is referred to the chapter on Epilepsy. In the cases of uræmic and other toxic convulsions, the treatment must be in accordance with the nature of the cause, and the condition is to be treated in every respect as the same condition would be treated if

it occurred in the adult. If the attack is supposed to be due to cutting teeth, the question arises whether incision of the gums is apt to be of any benefit. There has been much discussion on this point, and yet no definite conclusion has been reached; and no one, so far as I have been able to learn, has proved that cutting the gums has ever injured the child or impaired its future health. It is well enough, therefore, to try this method, and personally I have little doubt that a temporary relief of the hyperæmic condition of the gums is thus afforded. That the mere incision of the gum, as Henoch and Lewis suggest, is apt to increase the peripheral irritation, I can scarcely credit, if the incision be made according to modern surgical practice.

CHAPTER III.

EPILEPSY.

Few diseases have given rise to so much discussion as has epilepsy, the "morbus sacer" of ancient writers. Opinions regarding its cause and pathology have undergone many radical changes. The causes of epilepsy were by most writers thought to be obscure, and it is only within very recent years that a little more light has been shed upon the morbid processes underlying epilepsy. Some claim that epilepsy is never a disease *per se*, always a symptom, while others are inclined to regard true idiopathic epilepsy as a form of hereditary disease.

Whether epilepsy be merely a symptom or a distinct disease, the term may be used to describe a state in which convulsive seizures occur at varying intervals of time, and in which there is no special cause for each seizure. Thus a child may have a convulsive fit with the cutting of each tooth, yet we cannot say that it has epilepsy; but if that same child, after dentition and its attendant troubles have passed, continues to have fits at shorter or longer intervals, the child is surely a subject of epilepsy. Fortunately, however, only a very small proportion of those who have convulsions during childhood develop true epilepsy in later years. Webber reports that of 160 cases of epilepsy only 24 began before the age of five.

The Symptoms of epilepsy vary with the character of the attacks. Unfortunately the intervals between the attacks are not marked by as distinct a series of symptoms as is the case in tetany. In epilepsy the only symptoms that we can find in the interval are the effects of biting the tongue, the acne resulting from the bromide treatment, and the general stupor and indifference which are as often due to the drugs administered as to the disease itself.

Epileptic attacks can be divided into three classes:

1. Major attacks, or "*grand mal.*"
2. Minor attacks, or "*petit mal.*"
3. Epileptic equivalents.

Some patients are subject to major and minor attacks, and there is every possible gradation between the graver and lesser seizures.

The "grand mal" attacks differ but very little from the convulsive seizures as described in the preceding chapter. It will be sufficient in this connection to enumerate in the order of their importance and of their occurrence the chief symptoms of a major attack of epilepsy.

1. Prodromata, generally of a sensory character. At times there is a vasomotor or psychic disturbance.
2. Initial cry.
3. Loss of consciousness (very sudden).
4. Pupils dilated; no reaction; eyes open or closed.
5. Tonic, then clonic spasm of muscles (unilateral, partial, or general).
6. Spasm of respiratory muscles, which may lead to asphyxia.
7. Spasm of the muscles of the jaw (biting of the tongue, bloody foam).
8. Spasm relaxes, movements become clonic and then intermittent.
9. Involuntary passage of urine or of fæces.
10. Gradual recovery of consciousness, followed by a prolonged stupor or profound sleep.
11. The entire seizure lasts only a few minutes.
12. The deep reflexes are diminished or increased. The Babinski sign may be present.

In "grand mal" attacks most of these symptoms will be present, but occasionally we meet with major attacks in which there are no distinct prodromata; in which the initial cry is wanting; in which there is no biting of the tongue; no relaxation of the sphincters. The most constant symptoms are the loss of consciousness, the dilatation of the pupils, the spasm of the muscles, and the stupor or sleep after the convulsive movements have ceased. The diagnosis should rest upon the presence of at least two of these symptoms.

The prodromata are of great importance. In a fair proportion of cases the patient feels that an attack is coming on. A vague sensation at the stomach, a feeling of numbness or of tingling in any of the extremities, are by far the most frequent warning signs. In other cases the aura consists of slight twitching movements, of an altered psychic condition, varying from a general restlessness, or irritability of temper, to actual maniacal excitement. In many instances the character of the aura points to the involvement of a definite portion of the brain, and this is true even of cases of general epilepsy which are not supposed to be due to any gross cerebral lesion. In not a few cases temporary aphasia marks the coming on of an attack.

Auditory symptoms in the prodromal stage are not uncommon. In a little boy whom I described in a paper published some years ago, every epileptic attack was preceded by a hissing noise like that of a steam-engine letting off steam. A young lady, aged twenty-five, a school-teacher, who was subject to epileptic attacks at every menstrual period, is in awe of an old woman whom she sees regularly before each epileptic seizure. The vision of a ball of fire, of the colors of the rainbow, of a sudden change in the size of objects, is a common premonitory condition. Still another patient perceives a very foul odor during the aura; but I have never met with a case of epilepsy in which a pleasant or unpleasant taste preceded an epileptic fit. This would seem to show that the sense of taste is not independent of the sense of smell, although some authors refer in a vague manner to a gustatory aura.

Patients who have distinct auræ are on the whole more fortunate than those who have not, for they are able to prepare themselves for the attack. They can secure themselves against injury during the attack and may have time to apply remedies which either inhibit or shorten the spells.

The convulsive movements should be carefully studied in each case, for they help to define the character of the epilepsy. Generally speaking, universal convulsions point to hereditary epilepsy. Partial or unilateral epilepsy is indicative of organic cerebral disease. This distinction

would be a very easy one were it not for the fact that partial epilepsy may at any stage of the disease, and at any stage of an attack, become general, so that after the lapse of time the convulsions due to organic disease of the brain can in nowise be distinguished from those which are presumably hereditary and idiopathic. It is of the utmost importance, therefore, in making the diagnosis of epilepsy, to inquire into the past history of the case, and to determine, either by personal observation or by close questioning, the exact manner of onset of each attack. However rapidly the attack may become generalized, if it begins each time with distinct twitchings of the thumb, of the wrist, even of the eyelid, there is good reason to remove such a case from the category of hereditary epilepsy and to range it with those due to organic lesions. The attacks may come on at definite hours of the day or night. In "nocturnal" epilepsy an abrasion of the tongue, involuntary micturition, a tired feeling or a headache in the morning may be the only evidences of an attack.*

The typical "minor" attacks consist of a very transitory loss of consciousness, without any muscular twitchings, without the peculiar cry, and without the involuntary passage of urine and fæces. The loss of consciousness is often so slight that the child is supposed to have been "absent-minded" or merely "faint." Mothers and physicians, euphemistically inclined, are apt to speak of these attacks as "fainting spells." There is every reason to suspect epilepsy in any case in which "fainting spells" occur and recur without sufficient exciting cause. In some instances a sudden lull in the conversation of the child, a momentary stare, an unexpected stop when the child is practising upon an instrument, are the signs of petit mal; a change in the child's mental condition, in its character and temperament (there is often increased irritability) and sometimes a mild form of mania, reveal the serious character of what was supposed to be a mere fainting spell.

Bourneville has found a slight elevation of temperature during major attacks (not more than 1° F.); in the status epilepticus, the condition of continuous spasms, the tempera-

* For further details see description of convulsive attack.

ture may rise several degrees. Thomsen and Oppenheim have proved the existence of a concentric limitation of vision, and a diminution of general sensibility for some time after an epileptic attack.

The term "procursive epilepsy" has been applied to attacks in which the patient is suddenly impelled to run some distance forward, or backward; this may be a prodromal symptom, or it may be the only manifestation of an attack. This variety is very rare indeed.

We have alluded above to the existence of epileptic equivalents. Instead of having a typical convulsive seizure, the child passes into a state of mental confusion in which it becomes entirely irresponsible for, and unconscious of, its actions; it may also pass into a condition of trance or into an acute mania. Prolonged periods of double consciousness are rare in children; nor have I seen in very young persons the condition of narcolepsy (sudden falling asleep) which has been described as an occasional symptom of epilepsy in the adult. These "psychic equivalents" may be the beginning of a post-epileptic insanity.

The mental derangement following epileptic attacks is often marked by violent delirium. Under the influence of delusions and hallucinations, the patient may be impelled to deeds of cruelty. Such derangement may last for a few hours or weeks, or it may become chronic and lead to dementia.

CAUSES.—In considering the causes of epilepsy we shall take up, first, causes of the disease itself, and secondly, the causes which lead to individual attacks. That epilepsy is often a sad heritage can scarcely be doubted if we regard the statistics collected by Gowers, who finds a marked predisposition in two-thirds of 1,450 cases. The inheritance seemed to him to come more freely from the mother's than from the father's side. It is also beyond dispute that epilepsy in the ancestry is not the only predisposing cause. Other chronic nervous disorders have a powerful influence in this respect. I have known children of extremely hysterical, neurasthenic mothers to develop idiopathic epilepsy. Chorea in the mother is very apt to lead to the development of true epilepsy in the child; and, on the other hand,

syphilis and alcoholism in the father exert a very powerful influence in this same direction.*

According to Mendel's careful studies the majority of cases of hereditary epilepsy begin before the age of twenty years; but up to the age of forty years idiopathic epilepsy may manifest itself. The same author proposes to call the disease "late epilepsy," if it is developed after the age of forty years; in twenty-five per cent. of these "late" cases Mendel found distinct hereditary influences.

Accepting the truth of all these statements, I wish to impress one fact upon the mind of the student. Cases of hereditary (idiopathic) epilepsy are not nearly so frequent as they are supposed to be. If we examine carefully into the early history of our cases we shall find frequently that the child has either sustained some severe injury to the brain, or has acquired some cerebral lesion early in life. The paralysis and other symptoms which were due to the same lesion may have disappeared, but the epilepsy remains. I was led to this conclusion by my studies of infantile cerebral palsies, and my views have been quoted approvingly by various authors (among them Freud) who have become interested in this subject.

A very striking instance illustrative of this is that of a girl, about fifteen years old, who had been treated by many physicians for idiopathic epilepsy without any favorable result. On inquiring into the early history of the case I was told that several years previously the girl had suffered a slight paralytic stroke, every vestige of which had disappeared with the exception of an increase of the reflexes on one side of the body. Testing carefully for a possible diminution of power, I found distinct traces of an old hemiplegia. What was supposed to have been a case of hereditary epilepsy was clearly a case of epilepsy due to former cerebral disease.

The number of cases of *idiopathic* epilepsy will be still further diminished if we exclude from the number those cases in which there is evidence of defective general devel-

* Kowalewsky has insisted on the importance of syphilis in hereditary epilepsy, and has made a fanciful distinction between epilepsy due to paternal or maternal syphilis. His statements need corroboration.

opment of the brain; and those associated with infantile cerebral palsies; for the paralysis and the epilepsy are due to organic disease of the brain. The cases in which epilepsy is developed after an acute infectious disease cannot be considered "idiopathic."

The most characteristic cases of idiopathic epilepsy are those in which the disease is first noticed between the tenth and twentieth years. A few may come on between the twentieth and thirtieth, but those that occur later in life, or in the first few years of life, should be very carefully analyzed before the diagnosis of hereditary epilepsy is made.

It must be remembered that even in cases with distinct hereditary predisposition some other exciting cause must be present to develop the disease; thus we find that masturbation, disturbances of menstruation, sexual excesses, great emotional excitement, a blow to the head, are causes which lead to the development of epilepsy in those predisposed.*

Causes which Lead to Individual Attacks.—In a patient suffering from epilepsy any interference or disturbance of the general health of the patient may bring on an epileptic attack. Indigestion is perhaps the most powerful of these causes. Herter and Smith, in an excellent research on this subject, are inclined to look upon putrefactive processes in the intestine as a frequent exciting cause of epilepsy. They do not believe that this cause would be sufficient to produce epilepsy in a person not predisposed thereto. The same may be said of eye strain,† of urethral

* Reflex epilepsies are described by many authors as due to peripheral injuries (hand or foot); in such cases the aura begins with symptoms referable to the injured part. A permanent epilepsy is rarely established in this way, and I prefer to speak of reflex convulsions rather than of reflex epilepsy.

† Ranney's contributions to this subject are put forth very earnestly, but he fails to prove more than that the eye-conditions (errors of refraction, muscular insufficiencies, etc.), may cause single attacks; but they are surely not the cause of the epilepsy. Moreover, every neurologist knows that by treatment directed to the improvement of the eye-condition the attacks may be suspended temporarily, but the disease returns after a varying lapse of time. Ranney's cases are for the most part reported altogether too early. Hern and Dodd have given but little support to Ranney's (or rather Stevens's views), and their statistics are open to the same criticism. Instead of reporting the "cures" it would be valuable to publish the later history of all cases operated upon. In 1904 the author has again reviewed this subject and has seen no reason to change his views.

stricture, of narrow prepuces, and of laryngeal irritation. I doubt whether any one of these conditions has ever been the sole cause of epilepsy, though I am willing to concede that they may be sufficient to produce occasional attacks in persons with this special hereditary taint.

Improper oxygenation of the blood is another frequent cause of attacks. This explains why so many epileptics have their seizures in crowded court-rooms, in theatres, and in poorly ventilated bedrooms. The irritation of the epileptic centres in the lower portion of the brain is directly responsible for these attacks. After allowing for all these various causes we are compelled to admit that many of the epileptic seizures occur at more or less regular intervals, for reasons that we cannot discern, though I am firmly convinced that the more carefully we regulate the life and habits of the epileptic patient the fewer attacks he will have. It is very largely a question of removing all exciting causes.

A few powerful drugs have been known to cause epilepsy. Heiman described a number of cases of epilepsy due to poisoning by cocaine, and Tuczek has dwelt upon the potent influence of antipyrin. In the case of children cocaine need not be feared; but the careless exhibition of antipyrin might be responsible for unpleasant occurrences.

Gray has called attention to the frequent occurrence of epileptic attacks in those suffering from chorea and migraine. I have seen several patients, in advanced years, in whom the epilepsy followed upon the cessation of migraine.

Diagnosis.—The diagnosis of epilepsy offers but little difficulty if the character of the individual attacks has been clearly made out, and if such attacks recur at more or less regular intervals.

The epileptic nature of a seizure is determined by the presence of some of the several symptoms which go to make up a complete attack. The chief difficulty will be experienced in differentiating between an epileptic seizure and an ordinary fainting spell; also between epilepsy and hysterical attacks.

The characteristic features of each attack will be found in the following tables:

Epileptic Attacks.	Fainting Spells.
Loss of consciousness very sudden.	Loss of consciousness gradual.
Warning of short duration.	Warning of some minutes before consciousness is lost.
Pupils dilated; do not contract to light.	Pupils contracted or unaltered.
Pulse unaltered.	Pulse feeble.
Tonic and clonic spasms in various parts of the body.	No spasms.
Bloody foam at mouth.	No evidence of biting of the tongue.
Involuntary passage of urine and fæces.	No involuntary passage of urine or fæces excepting in rare instances.
Prolonged stupor after attack.	Recovery prompt after attack.

Epileptic Attacks.	Hysterical Attacks.
Loss of consciousness sudden and absolute.	Loss of consciousness not absolute.
Warning of short duration.	The attack often preceded by emotional excitement.
Pupils dilated.	Pupils not dilated.
Tonic and clonic spasm.	Tonic rigidity; exaggerated conscious movements; arching of back; excessive noises.
Eyes turned upward and inward.	Eyes staring, not turned, sometimes closed.
Involuntary passage of urine and fæces.	No involuntary passage of urine or fæces.
Prolonged stupor.	Recovery gradual; no stupor. The patient may pass, however, into a trance condition.
Attacks at rarer intervals.	Attacks may be frequently repeated.
Duration of attack short.	Duration of attack much longer.

The distinction between organic and idiopathic epilepsy can easily be made if we simply call to mind that organic epilepsies are, as a rule, partial, if not unilateral. Idiopathic epilepsies are invariably bilateral or general in their manifestations.

If organic disease is suspected, the manner of onset of the individual attack should be carefully determined, for partial

or unilateral manifestations often become general during the attack. Under the head of organic epilepsies we must include *post-hemiplegic* epilepsy. This form comes on with deplorable frequency after the paralytic attacks in early life. The epileptic movements as a rule affect the paralyzed side, but after the disease has lasted for some years the child, although its paralysis is unilateral, is prone to have general epileptic attacks. Under these circumstances the relation between the general epilepsy and the hemiplegic form of paralysis can scarcely be doubted. It will not be amiss to insist again upon the fact that the evidence of an existing hemiplegia may be so slight that it will be overlooked unless specially examined for; but however slight such traces may be, if it can be shown that the epilepsy was developed after the onset of the hemiplegia there is good reason to suppose that both the paralysis and the epilepsy are due to the same organic lesion. I consider it a safe rule in epilepsy beginning in childhood to examine particularly for exaggeration of the reflexes; and an increase of the reflexes in one-half of the body is quite as safe a sign of a preceding hemiplegia as a marked paralysis with contracture would be.

The sudden onset of epileptic movements in a child previously healthy should lead one to suspect the possibility of intra-cranial tumor; and a slight weakness of the parts convulsed, a possible increase of the deep reflexes in that same part, the presence of headaches and the development of optic neuritis, are the symptoms that we must look for in order to establish or to discard the diagnosis of tumor.

The same series of symptoms may occasionally be present in cases of acquired or hereditary syphilis; if so they are the result of general specific infiltration of the brain coverings, or of the development of gumma together with this general infiltration.

Prognosis.—The prognosis of epilepsy, whether of the organic or idiopathic order, is grave, not hopeless. Some patients with undoubted epilepsy have been cured. In many others, do what we will, the attacks recur as soon as treatment is abandoned. Painstaking treatment meets its due reward. The author's records contain the histories of

young men and women who were sorely afflicted in early life but have had no recurrence of epilepsy for periods varying from three to fifteen years. The attacks may be inhibited for a year and even for two years, but may return after that period of time if the customary treatment has been stopped. In this respect there does not seem to be a wide difference between organic and idiopathic epilepsy, except that in the former, malignant disease may lead to an early fatal termination. The frequent development of dementia, idiocy, or of epileptic insanity, makes the outlook gloomy enough in many instances. In spite of all recent therapeutic efforts we have not been able, however, to accomplish as much as is desirable in the way of a radical cure of the disease. The most encouraging hope that we have is that in the beginning of the disease the attacks may be due to special conditions which do not necessarily imply the existence of hereditary epilepsy. The fond hope that the child may outgrow the tendency to epileptic seizures is realized every now and then.

Pathological Anatomy.—Our knowledge of the morbid changes in epilepsy is very incomplete. Meynert laid great stress upon induration of the cornu amonis. Chronic hydrocephalus has been found in many cases, but this can hardly be considered a cause of epilepsy, for if it signifies anything it merely implies that the same process which led to the development of hydrocephalus also caused the epilepsy.

It is much more probable that the morbid changes of true epilepsy will be revealed through a study of the cortical changes which give rise to a localized epilepsy in cases of serious injury to the brain, and of epilepsy associated with infantile hemiplegia. In these conditions the initial stages of a morbid process* leading to epilepsy can best be studied; and it is possible, if not probable, that general epilepsy has such a beginning at a time and in a part of the brain unknown to us. Marie, Féré and Chaslin have led the way in these studies, and report the discovery of a neuroglia

* For other morbid processes associated with epilepsy see chapters on Cerebral Palsies and on Idiocy.

sclerosis which they consider to be the cause of epilepsy—such sclerosis starting from a focus of disease. These findings have been confirmed more recently by Alzheimer.

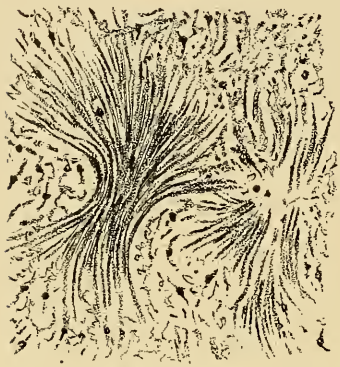


FIG. 41.—Diffuse Neuroglia Sclerosis of the Cortex in Epilepsy (after Chaslin).

Van Gieson examined cortical tissue, removed by McBurney, from two cases of Starr. He demonstrated most satisfactorily the changes in the large pyramidal cells of the cortex and in the neuroglia. He found that "the ganglion cells are affected by a series of degenerative changes, which in their most advanced stages result in an almost complete dissolution of the cell, and yet this degeneration is not extensive enough to involve the cells so universally as to interfere with their topographical distribution."

Some of the degenerative changes of the cells will be evident in the figure below taken from Van Gieson's article.

There is a decided change—a true hyperplasia—in the

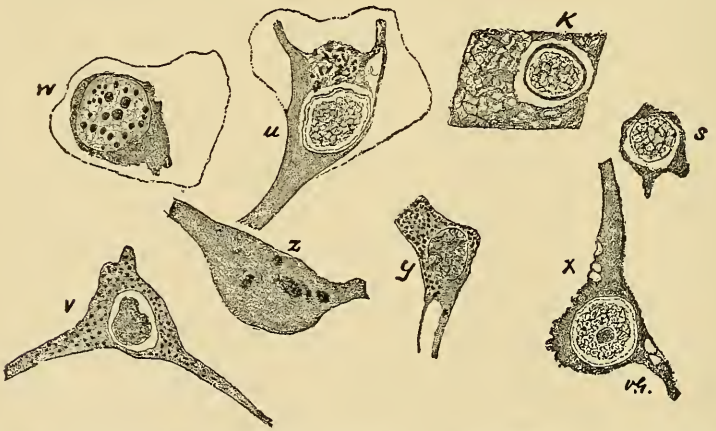


FIG. 42.—Various Phases of the Earlier Stages of the Degeneration of the Ganglion Cells. The thin lines enclosing the cells *w* and *u* represent the pericellular spaces; the cells *x* and *y* show the earliest stages, *w* and *s* later stages, and *k* shows the ultimate destruction of the whole of the ganglion-cell body, leaving nothing but the nucleus lying in an empty space. (Van Gieson.)

neuroglia tissue; clusters of an increased number of very young and seemingly proliferating neuroglia cells are visible in the vicinity of the small pyramids.

These findings were very much the same in two cases: in one the condition was due to a foreign body, and in the other an old cicatrix seems to have induced a similar pathological state in the neighboring tissue. To appreciate such researches at their true worth, it is important to note that they have reference to the early conditions only. What the nature of the secondary changes is, and how these are

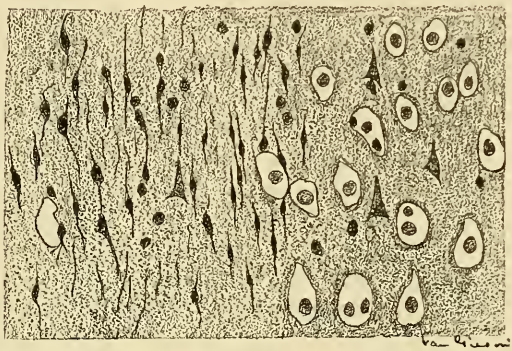


FIG. 43.—A Group of More Mature Neuroglia Cells in the Layer of Small Pyramids. (Van Gieson.)

developed from the original forms of disease is a subject for further study. Sailer has demonstrated the occurrence of an hypertrophic nodular gliosis in cases of epilepsy associated with dementia. Voisin questions whether this condition will be found in genuine epilepsy.

PATHOLOGY.—The pathology of the epileptic attack is identical with that of convulsive seizures in general. It is not necessary therefore to repeat in this chapter what was said in the chapter on convulsions. The only question of importance is whether in the diseased brain the convulsive centre on the floor of the fourth ventricle plays as important a *rôle* as it does in physiological experiments. Binswanger found that electrical stimulation of the lower half of the floor of the fourth ventricle in animals would produce tetanoid attacks or spasm in the limbs and the trunk, but he did not succeed in causing an ordinary epileptic at-

tack. There can be very little doubt that in man an epileptic attack is in the vast majority of cases the result of cortical irritation. The character of the sensory auræ is evidence of this, and so is the observation made by Oebeke and Gowers, that epileptic attacks cease after a lesion in the internal capsule.*

As for the pathology of the disease itself, opinions have undergone a very marked change within the last few years, owing to the rather startling suggestion of Pierre Marie, that epilepsy is often of infectious origin. Haig, Voisin, Féré and others advocate the theory of auto-intoxication, believing this to be due to the retention of toxic substances in the blood. Urine voided after an attack has been found to be more toxic than urine voided before the attack. Let us not forget that a focus of disease, due to purely mechanical causes, may be the beginning of general changes throughout the brain, which are responsible for the permanent establishment of epilepsy.

TREATMENT.—In discussing the treatment of epilepsy it is best to proceed on the theory that the attack is due to over-action of the cortical cells. Gowers explains the influence of the bromides by stating that "if we regard the morbid state in epilepsy as an instability in the resistance of the nerve-cells, it seems probable that the effect of the bromide is to increase the stability of that resistance." Whatever the explanation may be, we have up to this time found no drugs that can in any sense be considered proper substitutes for the bromides.†

It is certain that very few cases of epilepsy have been permanently cured by the administration of the bromides; but unquestionably they serve an admirable purpose in checking the number of attacks and in diminishing their

* Horsley pleads strongly for the invariable cortical origin of the epileptic attack, and believes that during the attack the cortex is in a congested rather than an anæmic condition. It will help us very little to say that the attacks are due to toxine poisoning. This may or may not be true; but in every case the most important question to settle is, why should certain individuals be prone to attacks and others not, while these same toxines from intestinal putrefaction, for instance, are present in hundreds of others who never have an epileptic seizure.

† A recent writer explains the action of the bromides by their power to diminish the effect of the toxines circulating in the body. If toxines and bromides were injected into the veins of an animal at one and the same time, the result was less severe than if the toxine alone was injected.

severity. To accomplish this end the bromide salts should be administered according to a definite plan. It has been my practice to give preference to the bromide of sodium, which I employ, according to the age of the patient, in ten or fifteen grain doses, three times a day. If given in a wineglassful of (alkaline) water after meals, the gastric functions will not be seriously impaired. Erlenmeyer suggested, some years ago, that a combination of the three salts—the bromide of sodium, the bromide of ammonium, and the bromide of potassium—would answer far better than the exhibition of any one of these salts singly. He went so far as to claim that if a single salt had produced acne, that that acne would disappear upon the administration of the three salts combined. I have given this method a fair trial, but have not been able to convince myself of the truth of Erlenmeyer's claim. The bromides should be pushed in every case to the point of tolerance, and until the attacks have been successfully diminished or inhibited. The loss of the palate reflex is evidence that the patient has become thoroughly brominized, but in children intolerance to the drug is very apt to be established long before this point has been reached.

The bromides can be administered in divided doses, two or three times a day; but if there is no good reason to give the drug during the day, there is a special advantage in giving the entire daily dose shortly before bedtime. The stupefying effect of the drug is not felt as distinctly as when it is administered during the daytime, and the sound sleep that ensues is an additional advantage to the patient. In the case of nocturnal attacks the medicine should be given before going to bed, and at no other time. In his various admirable lectures on the treatment of certain functional neuroses Seguin advocated the plan of giving the bromides on very much the same principle that one would adopt in giving quinine in malaria. In other words, the bromides should be given according to the periodicity of the attacks, giving little in the interval and increasing the dose very much at the time when an attack is expected; or if the attacks are frequently repeated, to give the drug four to five hours prior to the time at which the seizures occur.

I can subscribe thoroughly to this recommendation, as it has given me admirable results in the treatment of epileptic attacks in children. If the tendency to attacks has been overcome, the drug should be continued for a period of at least one year after the last attack. The dose may be diminished gradually, but it is wise to keep the patient in a state of mild brominism. The good effect of the drug can be maintained by giving, in conjunction with the bromide, five to ten grains of chloral hydrate, a few minims of the tincture of digitalis, or of the tincture of belladonna. Bromipin in daily doses of ten to twenty-five grammes has been useful.

Innumerable other drugs have been suggested; among these I will mention cannabis indica, which is particularly effective in cases of epilepsy with chronic headaches. The oxide and lactate of zinc have been freely recommended, but I have seen no beneficial results. The administration of iron and arsenic is thoroughly rational; no doubt the restorative effect of these substances upon the blood is the indirect cause of improvement in the epileptic condition. Some years ago Gowers advised the use of fifteen to thirty grains of borax several times daily. A fair trial has been given this drug in my clinic and in private practice, but I cannot claim more for it than for dozens of other methods.* Collins has pleaded in favor of nitroglycerin (in cases with a vaso-motor aura), but condemns simulo, osmic acid and hydrastis. Flechsig has warmly urged a combination of opium and bromides. Opium is given for six weeks; beginning with one-half to one grain; the dose is increased gradually until the patient takes eight, ten, or fifteen grains daily; then it is stopped suddenly, and the bromides in thirty-grain doses, four times daily, are substituted; after some time the dose is diminished to five or ten grains per day. I was ready to adopt this suggestion, as I had for many years been using codeine together with the bromide salts. In children the doses must be somewhat altered, but the plan of treatment, according to my own experience, deserves further trial.†

* Toulouse and Richet have advised the withdrawal of chlorides. Non-salted foods are to be given. The bromides will be more effective if less salt is taken into the system.

† Bechterew has advised a combination of bromides and Adonis vernalis.

The treatment of epilepsy is by no means exhausted by the recital of a few medicinal agents. Success depends upon other factors. The greatest possible attention should be paid to the details of the patient's daily life; regularity in the hours of sleep, in the hours of meals, and careful dieting, are essential to proper treatment.* I place my epileptic patients upon a general mixed diet, including small quantities of albuminoids and a liberal vegetable diet, but I forbid all indigestible articles of diet. I also limit the amount of cereals, and try to check intestinal fermentation as far as possible, being moved to this by the result of Herter's researches. Pastry and sweets of all sorts are strictly prohibited. Fresh air at all times, and particularly at night, should be insisted upon. Freedom from all emotional excitement is another recommendation that should not be overlooked. Marked ocular defects and other peripheral conditions which may cause an epileptic attack are to be treated.

Older patients who have distinct warning of their epileptic attacks should be provided either with the pearls of nitrite of amyl, which they can break up and inhale at short notice, or else they can be given a mixture of equal parts of chloroform and nitrite of amyl, which can be carried in a small phial and can be poured upon a handkerchief for purposes of inhalation as soon as the attack is signalled. In the case of partial epileptic attacks of definite onset a ligature applied around the part first convulsed, to be tightened quickly as soon as the warning comes, has been recommended by Hughlings Jackson, and is serviceable in some cases.

Surgical Treatment.—The surgical treatment of epilepsy has attracted great attention during the past fifteen years or more, ever since it has been known that the motor centres in the cortex can be safely and easily reached by the surgeon's knife. Surgical interference should be considered only in cases of partial epilepsy pointing to a definite focus of disease as the starting-point of the entire trouble. Nothing seems easier than to expose the centre

* In recent years rectal olive-oil injections, given at frequent intervals, have proved to be of great service.

of the part first convulsed in an epileptic attack, to remove this centre, and thus to remove the seat of disease, but, unfortunately, our ardent hopes in this matter have not been realized.

Some years ago Dr. Gerster and myself reported the results of operation in ten cases of epilepsy. The cases were carefully selected, and if surgical operation could have been of avail in any case of epilepsy, it would have been so in those we selected for operation; we were bound to admit that in our experience some slight improvement followed, but also that not a single absolute cure had been effected. Since that article was written my experience has been fully doubled, and the conclusions reached are practically the same. An analysis of the cases reported by Starr, which included my own cases, does not justify one in taking a more hopeful view of this question. I have referred in former writings to the reason for this disappointment. After an initial injury to the brain, months, and sometimes years, elapse before the epileptic habit is established. In about the same length of time a general cerebral sclerosis has been developed in connection with the original focus of disease. We may remove the original focus, but the general sclerosis remains and will act as a constant irritant upon the remaining epileptic centres.

On the other hand, it would be hasty to conclude that the surgeon's skill can be dispensed with in these cases.* The aim should be to watch carefully for the development of epilepsy, and to remove injured tissue at the earliest possible date. More than this, I consider it important not to await the actual development of epilepsy; and if the brain has sustained any considerable injury to remove the injured tissues, which, if allowed to remain, constitute a permanent menace to the future health of the patient. We shall be able to prevent the development of epilepsy very much more readily than we can cure it if once established.

The surgical procedures in vogue at the present day for the cure of epilepsy are trephining and excision of diseased tissues. Trephining has been practised for centuries. It

* The author wishes to direct particular attention to the excellent work done by Kocher.

is a method that entails no special danger upon the life of the patient operated upon, and can therefore be tried with perfect impunity except in very young children. White, of Philadelphia, and others believe that its only effect is the same as that of any operative procedure; but I am inclined to think that if it has any good effect it is in the way of relieving increased pressure, and that increased pressure is more frequently a real factor in epilepsy than is generally supposed, on account of the frequency of cystic formations and of the increased ventricular fluid in a very large proportion of the cases. Excision of tissue would seem to be a rational procedure in the earlier stages of epilepsy developed after skull injury, or in connection with early cerebral disease; but the paralysis which so easily results from the excision of the motor centres militates somewhat against the advisability of this practice, although according to Dana's and my own experience such paralysis disappears after a few weeks. In cases in which the epileptic attack is preceded by sensory or psychic phenomena, a removal of the auditory or visual centres, for example, would be a serious matter indeed, if not entirely unjustifiable. Many a patient would prefer the occasional repetition of an epileptic attack to a permanent loss of hearing or a permanent impairment of sight.

The only other* surgical procedure which has been suggested for the cure of epilepsy is the ligature of one or both vertebrals, as was first done by Dr. Alexander, of Liverpool. The dangers of this operation are so great, and the results so uncertain, that it scarcely merits serious consideration.

* I do not consider it necessary to refer to the surgical treatment of the eye-muscles for the *cure* of epilepsy except utterly to condemn the practice. As I am treating of epilepsy in children, I need not discuss oöphorectomy and similar procedures.

CHAPTER IV.

HYSTERIA.

TRUE hysteria is a relatively rare condition in adults.* This may seem a very heterodox statement to those who have been ready to apply the term hysteria to many affections of the nervous system which could not be attributed to organic lesions. Those who are advised of the recent conception of hysteria will not hesitate to indorse the author's opinion. The determination of the chief symptoms of true hysteria, and the separation of this disease from many other functional disturbances which were once classed with it, have constituted a signal achievement in modern neurology. We have at last succeeded in establishing a number of symptoms, chiefly of a sensory character, which, when present, enable one to make the diagnosis of hysteria with a great degree of certainty, and which do away with the necessity of reaching this diagnosis by the process of exclusion. As the physician's knowledge of nervous diseases increases, the diagnosis of hysteria is made more and more infrequently. In the author's very large clinical service at the New York Polyclinic the diagnosis of hysteria was one of the rarest of all diagnoses made. In other institutions with which the writer is connected, particularly in the Montefiore Home for Chronic Invalids, hysteria is found to be much more frequent, especially among the foreign-born inmates. If hysteria is a rare disease in the adult, it is still rarer in the child, but when it does occur it is an important factor in the future life of the child. Jolly, Ferrier, Oppenheim, and others report cases of hysteria occurring as early as the second or third year of life.

* We see much less of true hysteria in this country than in Europe; it is also far less common in England and in Germany than in France and Russia. Broadly speaking, the Anglo-Saxon race is less prone to the development of hysteria than the other races represented in our population.

In the development of hysteria, hereditary influences are the most potent factors. Charcot and his followers rule out all other causes. Given an hereditary predisposition, mental overexertion, fright, emotional excitement, exhausting diseases, trauma, masturbation, early sexual irritability (Freud) may be the contributing causes.

English and American authors have had very little to say upon the subject. In the large hand-book of Gerhardt, Jolly has treated the subject in a very satisfactory manner, and in American Cyclopædias and Systems of Medicine the subject has received careful attention at the hands of Mills and Lloyd. Henoch has written a very full chapter on the subject, but includes under this heading many forms of disease which are more properly designated by other names. The French authors have naturally had much to say on the subject, and they include so many different forms under this heading that one is puzzled to know whether there is any form of nervous or mental disease that is not in some way related to hysteria or influenced by it.

The symptoms of the disease are truly protean in character. No one person, fortunately, ever exhibits even a majority of them. Hysteria in one person is very different from the disease as it is exhibited in others. It is difficult, therefore, to give any one clinical picture of the entire disease. It will be best to take up the chief symptoms as they occur independently of other diseases, or as they are known to complicate other functional and organic diseases of the nervous system. Hysteria is characterized by symptoms which point to a defect in the various systems and organs of the body. Thus we have:

1. Psychic manifestations.
2. Motor manifestations.
3. Sensory manifestations and vasomotor disturbances.

1. PSYCHIC OR MENTAL HYSTERIA.—Properly speaking, every case of hysteria would come under this heading. For, if there is any one feature that distinguishes hysteria from other diseases, it is the defect of will-power and the excess of the emotional faculties. "I can't" is the pet phrase of all hysterical subjects, or still better, "I will not." It is not so much a direct lack of power to exert the will,

as a tendency to exert it in perverse fashion. This peculiar mental condition is easily recognized, and often leads to the diagnosis of hysteria in persons who have other symptoms pointing to a much more serious disease. But aside from this general hysterical state of mind there are other mental conditions which are very typical of hysteria. The most pronounced psychic form of hysteria, as observed in children, is that known as *hysterical mania*. Under great mental strain or excitement a child is seized with a crying or laughing spell, after which it passes into a state of nervous excitement in which, as in a little girl, aged eleven, under my observation, the child becomes violent, attempts to strike others, to injure herself, and to tear her clothes from the body, and to do all possible injury irrespective of consequences.

Another condition, sometimes classed under the heading of hysteria, is observed in young girls, particularly at the age of puberty, and often ends in a condition of marked acute mania. I have seen a number of such cases in girls who were over-ambitious and eager to pass their school examinations. They would keep up under the excitement of the examination, but immediately thereafter, whether successful or not, would become irritable, excitable, sleepless, would have laughing and crying spells by turns, would refuse to take nourishment, and eventually would either recover under proper treatment, or else pass into a condition of typical acute mania with absolute loss of reason, with intense excitement, and confused delirium. There would be no reason to consider these cases in this connection if they did not occur in children who have shown either a marked nervous predisposition, or who have exhibited hysterical symptoms of one kind or another at previous periods. Such children, if subjected to severe strain or severe emotional excitement, are very apt to pass into a condition of hysterical mania.

Other mental conditions which cannot well be separated from hysteria, are those known as hystero-epilepsy, catalepsy, hysterical trance, and the like. Of these, hystero-epilepsy is by far the most important, and the gravest disorder. Mills is inclined to consider this form of disease

very rare among children. In this he is undoubtedly correct; but it has been my good fortune to see a number of classical instances of this special form in children, and the account I give is based entirely upon personal impressions. The gravity of hystero-epilepsy is increased by the fact that the children so afflicted are found most frequently in degenerate families. Insanity, epilepsy, chorea, chronic alcoholism, are the predisposing conditions in the ancestors of those who suffer from hystero-epilepsy. In one family I have during a period of ten years treated the mother for severe hysteria, a daughter at the age of nine for hysterical convulsions, and another son and daughter for typical hystero-epilepsy.

On account of this close relationship to true epilepsy, cases of hystero-epilepsy cannot be regarded with the indifference which many physicians still display toward hysterical subjects. In many cases it is difficult to decide whether the attacks as they occur are more hysterical or more epileptic; and of the attacks occurring in one individual some may be typically hysterical, while others may be typically epileptic; and there is some danger in every case of severe hystero-epilepsy that with the progress of years the hysterical symptoms may vanish and true epileptic attacks may occur. It is of the utmost importance, before passing judgment upon any case, to determine whether the attacks are epileptic or hystero-epileptic.

2. Motor Manifestations.—Every variety of clonic and tonic movements occur in hysterical subjects. Many of these will be understood best by referring in detail to a few cases of typical hystero-epilepsy.

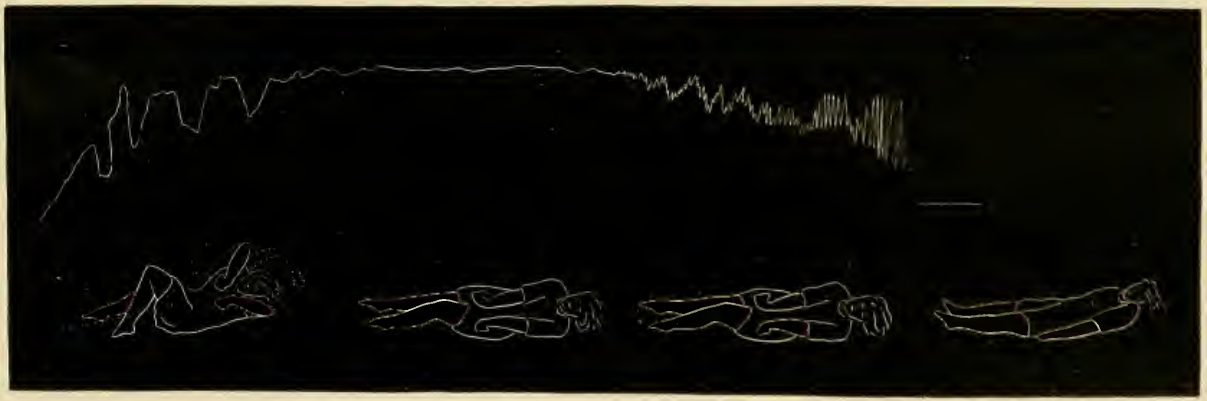
Some seven years ago I was called to see a lad, then thirteen years old, who had been seized with violent convulsions during the night. I had been informed that these convulsions were preceded by great emotional excitement, caused by a severe upbraiding which the boy received for his misbehavior. After this little domestic scene was over the boy went to bed, and was seized with the first attack. During the attack he gave a shrill cry; then began to bark like a dog, snapping at everyone who approached him, and would pass through the most severe contortions, touching

the bed at times only with the head and heels, the back being deeply arched as in the famous drawings of Richer representing this condition in women. After this the most violent jactations of the entire body occurred. During these convulsive movements he would snap, bark, and bite, then he would suddenly give a leap to the foot of the bed—almost tumbling out of bed—yet he always saved himself in time. This whole performance would last about two or two and a half minutes, then thorough relaxation of all the muscles would take place, he would fall back exhausted, and would then regain consciousness. During the attack there was no evidence of consciousness, at least no impression could be made upon him through any of the special senses. The patient often had as many as two hundred such attacks in the course of twenty-four hours; he was sleepless, refused food, and became very much emaciated. After careful nursing for a period of six weeks the attacks gradually lessened, and eventually he recovered entirely; but he has shown since a deficiency in his moral and mental make-up, and although he has been cured of the hystero-epilepsy, it was found necessary to place him in a reformatory. This last fact is of some interest as showing the relation which hystero-epilepsy bears to degeneration of all the mental and moral faculties.

Another case was that of a young girl, aged fourteen, who was much worried over the fear that she and the family would starve, as the father had daily drummed into the child's ears the necessity of economy and the difficulties of supporting a family in hard times. The young girl, the daughter of a very hysterical and emotional mother, took the warning to heart, and for the first time in her life developed typical hystero-epileptic attacks. Without warning she would fall from a chair, from a sofa, or even on the street would pass suddenly into a condition of apparent unconsciousness. Wherever she lay she would pass through all sorts of contortions, would shriek, now and then would give agonizing yells, and would finally end up by a pronounced crying spell. At other times she would become violent, tearing her own clothes, the bed-linen, and everything that was within her reach in the room. These spells

FIG. 44.—Epileptoid Stage of Hystero-Epileptic Attack. *Tracings.* (Richer.)

1. Tonic Stage. 2. Clonic Stage. 3. Relaxation.



Tonic Convulsions. Tetanic State. Clonic Convulsions. Stertorous Sleep.

(From Grasset et Rauzier.)

were repeated very frequently during a period of three weeks, and then gradually subsided. During all these attacks the movements seemed to be more or less purposive, and yet there was good reason to think, and the girl later on confessed, that she was absolutely unconscious of what she was doing at the time. These two cases will suffice as a general indication of the common form of hystero-epileptic attacks.

The distinction between the attacks of epilepsy and of hystero-epilepsy is brought out in the following table:

EPILEPSY.	HYSTERO-EPILEPSY.
Aura frequent.	No aura; but,
No distinct cause for each attack.	Some emotional excitement, direct cause of attack.
Onset sudden.	Onset sudden or gradual.
Initial cry.	Noises of all sorts during attack.
Eyes open or closed; pupils often dilated widely; do not react; rolling of eyes upward and inward.	Eyes turned up; pupils normal; often ecstatic expression of countenance.
Tonic and clonic convulsions.	Either tonic rigidity of muscles or extravagant movements (sometimes purposive).
Involuntary micturition or defecation.	No impairment of vesical and rectal reflexes.
Duration of attack only a few minutes.	Attacks last much longer, followed by a condition of trance, or else patient recovers consciousness as soon as convulsive movements cease.
The patient often injures himself.	The patient falls softly, without personal injury.
Biting of the tongue.	Biting of the tongue rare.
After cessation of clonic movements, stupor or somnolence.	Patient may pass into condition of trance, or may exhibit signs of fatigue; recovery often rapid.

In addition to the typical attacks of hystero-epilepsy there are other forms of motor disturbance due to hysteria. The hystero-epileptic attack, while it is the gravest, is in fact much rarer than the ordinary hysterical convulsion. The latter is characterized by a temporary and imperfect loss of consciousness; by irregular, though sometimes violent, twitchings of all extremities, sometimes by the repetition

of one special form of convulsive movement, such as retraction of the head. When these various convulsive movements are over, the muscles pass into a state of tonic rigidity, after which rapid recovery takes place. The entire episode is wound up by a fit of crying or laughing, or by some other expression of an emotional character. Some of the patients pass into a condition of melancholy which may last for hours, or into a state of exaltation and even of ecstasy. This is as true of the hysterical attacks as they occur in children as it is of those that occur in the adult. These attacks can be distinguished from the hystero-epileptic and from epileptic seizures by the incomplete loss of consciousness, by the absence of all regular rhythmical convulsive movements, and by the fact that there is neither biting of the tongue, nor involuntary micturition, nor any of those symptoms which are often associated with hystero-epileptic and epileptic attacks. Other hysterical attacks consist of spasm of the head and neck muscles, the well-known spasm of the œsophagus which gives rise to the sensation known as globus hystericus, or to difficulties in deglutition if the spasm affects the lower part of the gullet. This œsophageal spasm which occurs in hysterical and otherwise neurotic children is a symptom which has not met with the appreciation which it deserves. If a sound is passed into such a gullet it will be seen that there is an actual muscular spasm at the point of constriction, and that it requires considerable patience, and sometimes a little force, to overcome the contraction; but the age of the child and the fact that the constriction easily disappears, to return again under the slightest emotional excitement, is sufficient to remove any fear of the constriction being due to an organic cause. In this same category we may place spasmodic movements of the diaphragm, of which singultus is the common manifestation. This special phenomenon is much more frequent in later periods of life than during the period of childhood; but I have had under observation an entire family afflicted with this special form of hysteria who upon the slightest provocation would exhibit this symptom. On one occasion I examined the mother, in the presence of two daughters, respectively twelve and ten

years of age. Under the excitement of the examination the mother was seized with severe singultus, and before I had completed the examination both the daughters were favoring me with a similar exhibition. The scene would have been a comical one if it had not brought home to me the powerful influence of example and suggestion, if not of heredity, in such cases.

Spasm of the bladder, increased peristaltic action of the stomach and bowels resulting in diarrhœa, upon the least emotional excitement, fits of crying or laughing, of sneezing and of coughing, are the various forms of hysterical spasm met with in children, as well as in persons of more advanced age. If the hysterical seizure, or better said, the hysterical spasms, assume a more extravagant character, and if it affects a larger number of the groups of muscles that generally act in unison with one another, we may have that peculiar condition which is known as *chorea major*. In this form the jactations are often severe and exhausting. The child or young person assumes the most impossible positions, often suggesting the intention of dramatic effect. From this condition it is but a short step to a condition of mild tonic contracture, the limbs retaining any position which may be given them. This state of catalepsy is very often witnessed in conjunction with chorea major, sometimes independently of it, but it is a mistake to assume that catalepsy occurs exclusively in conjunction with hysteria. It occurs in connection with various forms of mental derangement, and a condition not unlike catalepsy is met with in some forms of infantile cerebral paralysis.

In hysteria the muscles of the body are not only subject to convulsive movements but also to absolute paralysis. Every part of the body, including the ocular muscles, may be the seat of such palsy, but it is most frequently manifested in the extremities, in the tongue, and in the vocal cords (hysterical aphonia). In some instances there is no absolute paralysis of any one muscle or group of muscles, but certain functions are paralyzed. The best known example of this is the form which French neurologists have termed astasia-abasia. A person thus afflicted is neither able to

stand nor to walk, but is perfectly well able to use all muscles while lying on the back. Many have claimed that this is simply due to a psychic condition dominated by the idea that standing or walking is impossible. This is in all probability the proper interpretation; but the point that concerns us here is that it rarely, if ever, occurs except in those who exhibit other symptoms of hysteria.

Hysterical aphonia is very frequent in children, and particularly during the period of development. The hysterical character of the aphonia is easily recognized by the fact that (as the laryngoscope reveals) there is no organic cause for the loss of voice, and that the single attacks come on very suddenly, as a rule, in the wake of some nervous excitement, and disappear as rapidly as they have appeared. I have had young girls come to my consulting-room who would not be able to speak above a whisper, and it has always been a special pleasure to have them sing the entire scale before leaving the room. A strong faradic brush applied to the neck over the trachea is the most persuasive master in these cases. Hysterical mutism, an absolute loss of speech, is generally the result of severe emotional excitement. In some instances it is associated with delusions, and with other symptoms of mental derangement.

Other forms of hysterical paralysis, ocular palsies, for instance, are not easy to diagnosticate, and in order to distinguish them from paralysis due to organic causes the physician must have all neurological facts and doctrines at his command. It is indeed one of the most difficult problems in neurology to distinguish hysterical from organic forms of paralysis, but this distinction can be made safely enough if the following points are kept in mind: Hysterical paralysis does not as a rule adhere to anatomical distribution, with the one exception that an hysterical hemiplegia may be quite as complete as any hemiplegia due to an organic disease. But I have not yet met with a single case of hysterical hemiplegia in a child that has in any way suggested, even for a moment, the possibility of a hemiplegia due to an organic lesion in the brain. In hysterical paralysis the paralysis, as well as the anæsthesia which is associated with it, is apt to be regional; thus we have a paralysis of the hand, or a pa-

ralysis of hand and forearm, or paralysis of an entire extremity, with an anæsthesia that, as a rule, develops in proportion to the paralysis. This association of regional anæsthesia and regional paralysis is, to my mind, very characteristic of hysteria. Hysterical contractures are not infrequent.

From cerebral palsy, hysterical paralysis can be distinguished by the fact that it is not accompanied by increased reflexes; and by the lack of marked sensory changes in the majority of cases of brain paralysis. The deep reflexes may be lively in cases of hysterical paralysis, but I have never seen them markedly exaggerated, nor are they accompanied by the spasticity and tonic contractures which are of such frequent occurrence in the case of paralysis due to brain disease. The presence of ankle clonus was at one time supposed to prove the non-hysterical character of an affection; this may be true in the majority of cases, but since I have met with the presence of ankle clonus in some hysterical, and even in neurasthenic, affections, I cannot attach much importance to this one point. The electrical reactions, it should be remembered, are normal both in hysterical and in cerebral diseases; we cannot therefore utilize them in any way in making the differential diagnosis.

If the question arises whether paralysis of the upper or lower extremities (more frequently of the latter) is due to spinal disease, or whether it be purely hysterical, the diagnosis should be based upon the absence of vesical and rectal symptoms in a case of hysterical paralysis, though to be sure there are many cases of spinal disease too in which these are not affected. Retention of urine may occur in cases of hysteria, but it is only of short duration as a rule, and is rarely accompanied by the symptoms of cystitis, as is the case in a large number of spinal palsies. In many cases of spinal and peripheral nerve disease the electrical reactions are altered, and the reaction of degeneration may be present. This is not the case in hysteria; but these electrical reactions are normal also in all those cases of spinal disease which are due to disease of the lateral columns and not of the anterior or central gray matter.

All these points of differential diagnosis may in some cases prove insufficient, and the diagnosis of hysteria or of

organic disease of the nervous system will depend upon the general agreement or disagreement of symptoms. If the physician is aware that flaccid paralysis of a single group of muscles, or of one or more extremities, is generally associated with changes in the electrical reactions, and with loss of reflexes in case these symptoms are due to organic disease, and if he remembers, furthermore, that spastic forms of paralysis are associated with increased reflexes, with normal electrical reactions, and with normal sensations, he will have little or no difficulty in arriving at a correct diagnosis. Hysterical paralysis is subject, moreover, to greater changes than the organic palsies are. While under certain conditions the very persistence of the symptoms for a long period of time, followed by a sudden change, is extremely characteristic of hysterical paralysis, the diagnosis becomes still more certain if the hysterical paralysis is associated with other symptoms, such as are known to be purely hysterical. I refer particularly to the presence of rigidity, anæsthesia, or typical and complete hemianæsthesia, and to the occurrence of major or minor hysterical attacks; but the difficulties are still further increased by a fact to which Seguin called particular attention some years ago, that hysterical paralysis or hysterical symptoms often complicate organic disease.

French authors have reported a number of cases of extreme progressive wasting of muscles in hysteria. Hirt has recorded a case in a young girl, so astounding in the rapidity and degree of development of the atrophy that it almost challenges belief.

3. Sensory Symptoms.—From the preceding pages it is evident that the sensory symptoms often enable one to make a positive diagnosis of the hysterical character of the affection when other symptoms would leave some room for doubt. Every form of sensation may be affected, and it may either be exaggerated or dimished. Hysterical hyperæsthesia and hysterical anæsthesia are of common occurrence. If hyperæsthesia exist there is an unusual sensitiveness to the slightest touch or to the slightest painful impression. This hyperæsthesia is most prominently developed in the region of the ovaries, in the skin over the

spinal column, each spinous process being so sensitive to touch that the patient cries out from pain, however light the touch may be. The hyperæsthetic areas, if stimulated, are very apt to cause distinct hysterical attacks in those prone to such seizures, and the hyperæsthetic areas may become true hysterogenic zones. These hysterogenic zones have been studied with great care by the French school, but we cannot enter upon the subject more fully here, and leave it with the simple statement that there are some hysterogenic zones which are not hyperæsthetic.

Hysterical anæsthesia is still more frequent than the exaggerations of sensory impressions. The anæsthesia may not only include every form of ordinary sensory perception but also the special senses. The distribution of this anæsthesia is one of the most valuable signs of hysteria. It occurs in the form of a complete hemianæsthesia, or in the form of a regional anæsthesia. The hemianæsthesia is limited strictly to one-half of the body, but in this half it is often absolute, and the completeness of the anæsthesia is the very feature which should make us suspicious of its hysterical nature. If a girl or a boy, or for that matter any person at any age, present absolute anæsthesia to all forms of ordinary sensation, and in addition exhibits blindness of one side or a typical hemianopsia, deafness, loss of taste, and of smell in the same half of the body, such a person has undoubtedly hysteria. In this country hysterical hemianæsthesia is relatively rare, excepting in the Russian and French elements of our population, but in these it can be studied easily and satisfactorily.

If an entire half of the body is not anæsthetic, the regional character of an anæsthesia is sufficient to lead one to the diagnosis of hysteria. By regional anæsthesia I mean anæsthesia of a well-marked division of the body, say of the hand, of the forearm, of the leg, the hips, or of circumscribed areas in any part of the body without reference to the anatomical distribution of the sensory nerves. (Fig. 45.) As this regional anæsthesia is frequently associated with a regional paralysis the hysterical nature of both becomes very evident. The hysterical nature of the anæsthesia is not only made evident by its peculiar manner of develop-

ment, but also by the manner in which it often disappears. In a number of cases in which I have studied this anæsthesia carefully it would recede inch by inch, but always maintained a certain level in the arms or legs without any reference to the well-known sensory areas. Hysterical patients do not present anæsthesia of the anterior or posterior surface of a limb, but the anæsthesia generally involves both surfaces. The hysterical nature of sensory disturbances is also established by the powerful effect of hypnot-

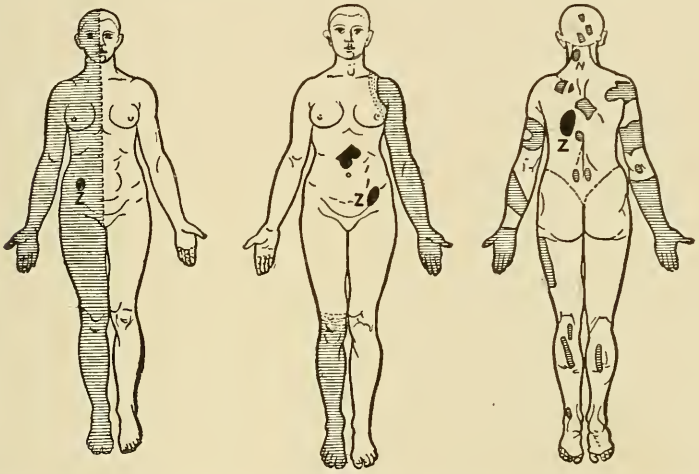


FIG. 45.—The Three Types of Distribution of Anæsthesia in Hysteria: Hemianæsthesia, Segmental, and Disseminated. Z, Hysterogenic Zones. (Dana.)

ism, and of mere suggestion, which is often sufficient at least temporarily to dislodge a complete anæsthesia. The anæsthesia may furthermore be transferred from one limb to another, not merely by the action of magnets, or by the action of metals as was formerly supposed, but, I am convinced, by any form of suggestion powerful enough to produce a strong psychic impression. It is on the theory of suggestion that we must explain the miracles ascribed to magneto-therapy or metallo-therapy, that played such an important *rôle* about twenty-five years ago in France.

Of the special senses, vision is most frequently subject to hysterical disorder. In some there is true photophobia, in others a diminution of visual perception is more common,

and the patient may indeed be unaware of the existence of such diminished visual sensation, which is the best proof that it is not intentional or simulated. The retina may be entirely insensitive to light, there may be distinct limitation of the visual field, or there may be a complete loss of every form of visual perception in one eye. (Fig. 44.) Bilateral loss of sight is said to occur in hysterical patients, but as a rule it is simply transitory. I have not had opportunity to see this special visual defect in children.

Visceral hysteria deserves a passing notice; the paralysis of the bladder and increased peristaltic action of the

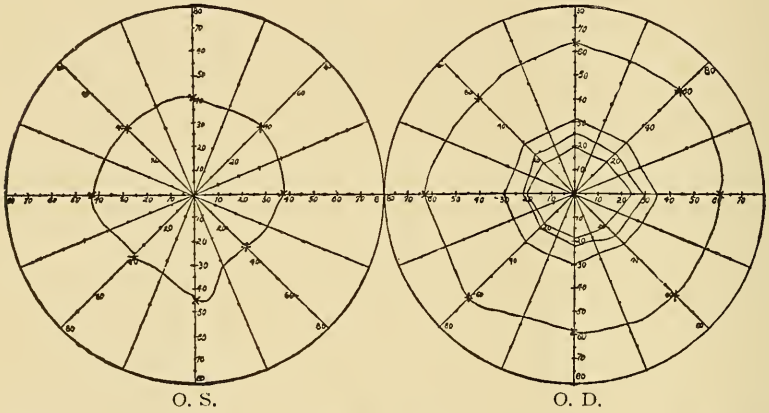


FIG. 46.—Hysterical Loss of Color Sense and Limitation of Visual Field. Color Sense Absent in Left Eye and Field Contracted; in Right Eye Field less Contracted; Order of Appreciation of Colors from Without In was Yellow, Violet, Blue, Red, Green. (After Peck, from Dana.)

stomach and bowels have been mentioned in connection with the motor disturbances of hysteria, but there are two other forms of hysterical manifestations that are worthy of further mention. The one form is hysterical anorexia. Patients in this condition absolutely refuse food, and if they take it, vomit it at once. In some instances this is accomplished without effort on the part of the patient and seems entirely unintentional. In other instances, again, the patient deliberately sets to work to eject the contents of the stomach, and does not give up the effort until she has succeeded. In my service at the Montefiore Home I frequent-

ly had occasion to witness the antics of an hysterical girl, aged fifteen, who immediately upon taking food would go into a corner of the room, begin violent gagging movements, and would succeed within a period of three or four minutes in ejecting the entire contents of the stomach. In this girl, who was not possessed of any insane delusion regarding the character of the food, the act seemed purely volitional, and it was a surprising fact that like many other hysterical patients with anorexia, she was not so much reduced in her physical condition as a person would have been who had persistently starved herself. She evidently obtained food and retained it; but how she got it and when she took it it was difficult to determine, since she was watched carefully night and day. She was treated by rather heroic measures in the form of forced feeding and hydro-therapeutic measures; she was allowed to leave the institution after she had shown her willingness to take food and retain it.

Distention of the stomach and bowels, representing a total paralysis of the muscular walls of the stomach and intestines, occurs frequently enough, and more particularly in children. The stomach is apt to be distended to an enormous degree, and the bowels dilate in such a way as to give the appearance of the abdomen in a case of large-sized tumor. I have in mind the cases of twin brothers who presented the following peculiar condition: The one brother, at the age of fourteen, was true to the neurotic stock from which he descended by suddenly developing a tremendous meteorismus, which increased from day to day until the abdomen was so distended that the skin seemed ready to burst. There was some anorexia and constipation. The constipation was occasionally relieved, with the result of temporarily diminishing the distention of the bowels, but in a very few hours the intestines would resume their former state. Every possible measure was resorted to, to remove this distention, but it persisted stubbornly for some weeks, when finally, after the introduction of large enemata of an infusion of valerian, the distended abdomen suddenly collapsed. A few days later the twin brother of this patient began the same performance, and went through exactly the

same experiences. The abdomen became distended to almost the same degree as in the brother, and after it had lasted about the same length of time his abdomen also collapsed in very much the same way. These boys not only came from very neurotic stock, but had been very much pampered in their early training, which fertilized the soil that had been prepared for the development of every form of hysteria.

Diagnosis.—It would be impossible to state the points of differential diagnosis between hysteria and other affections without repeating everything that has been said in the preceding paragraphs. It is worth while stating once more that the diagnosis of hysteria should be made only in case an organic affection can be positively excluded, and if the well-recognized symptoms of hysteria, particularly hysterical seizures and hysterical sensory changes are present. Let the student also bear in mind that hysteria and hypochondriasis are not one and the same disease, though one is often mistaken for the other. If a boy is fearful of a disease with which he supposes he is afflicted, we have no hesitation in saying that he is a hypochondriac; but let a girl exhibit exactly the same symptom, and she is at once put down as an hysterical subject. To distinguish between the two conditions is not always an easy matter, but in hypochondriasis the patient is generally possessed of a few notions regarding his own bodily condition, and these make such a deep impression upon him that his entire ego is affected by it. His supposed affliction is constantly before his mind, and dominates his entire being. In cases of hysteria there is no such introspection as in hypochondriasis. The bodily ailments or bodily peculiarities are far more numerous, but they are subject to greater changes, disappear for a time and then return again, and are not pushed to the fore quite as much as in cases of hypochondriasis. In the latter, moreover, the depression is, as a rule, greater than in hysteria, and in hysteria there are other symptoms which prove the presence of an hysterical affection. The difficulties of a differential diagnosis are still further increased by the fact that a mixture of the two conditions (hysterical hypochondriasis) is not uncommon, and it must be left to the physician to determine, by a close analysis

of the symptoms, whether there is more hysteria or more hypochondriasis in the symptoms which the patient presents.

PATHOLOGY.—In discussing the pathology of hysteria there is much room for theorizing, but there are very few facts to guide us. That it is a functional disease is conceded on all sides; but there is no other disease in which the loss of function may be so absolute as in hysteria. By some mechanism or by some influence which we cannot yet understand, an entire hemisphere is temporarily invalidated, or else we could not explain the typical and complete hemianæsthesia so common in hysteria. Meynert endeavored to give an anatomical explanation, but failed. How this loss of function of one hemisphere is effected, and whether it is similar or not to the occurrences that take place under hypnotic influence, we are not prepared to say. The transfer of sensory disturbances from one-half of the body to the other, would go to show that the two halves of the brain are evidently in sympathy with one another (to put it broadly) in this disease, and that they can be alternately affected. The highest centres are evidently impaired in hysteria, cortical inhibition is removed, and the lower centres have full sway.* It is quite in keeping with this view that even the reflexes are frequently exaggerated in hysteria as they are in organic disease in which the changes in the motor fibres of the pyramidal tract interfere with the proper transmission of cortical influences. We cannot properly speak of the morbid anatomy of hysteria, for the entire conception of the disease would have to be altered if we could suppose the existence of such. Some post-mortem findings have been reported, but they were evidently accidental complications.

DURATION AND COURSE.—Once hysterical always hysterical, would seem to be the general opinion with regard to these subjects, but this is not quite accurate. The manifestations of this disease often disappear for years, and it is

* Breuer and Freud have attempted a psychological explanation of hysterical phenomena and have ascribed these to a "hypnoid" condition in which there is a division of consciousness, and a limitation of the power of association. Freud has directed attention to the diminution in the strength of concepts in hysterical subjects, and to the ready change from psychic to physical innervation.

one of the commonest experiences to record the disappearance of an hysterical paralysis or of an hysterical anæsthesia. The tendency to relapses is extremely great, and symptoms that have disappeared for years may occur under any emotional excitement.

TREATMENT.—The treatment of hysteria must be considered with reference to the hysterial predisposition and with reference to the special hysterical symptoms. The hysterical predisposition is unfortunately either inherited from a neurotic ancestry or it has been specially fostered by an hysterical environment. In hysteria, as in other mental affections, I have often doubted whether heredity is the marked factor that it is generally supposed to be, or, if we grant the hereditary tendency, whether the hysterical manifestations would be as frequent as they are if a serious attempt were made to change the surroundings of such children. In the majority of cases the early training has been defective, and an example constantly before the child of an hysterical mother or other hysterical relatives is sufficient to engender the disease in its fullest form. The first and most important principle of treatment, therefore, is the absolute separation of the child from the family. It is a great pity that this is so rarely urged by the physician, and still more rarely permitted by the parents. Only the more intelligent parents can be made to understand that an utter stranger, if properly qualified, may train a child far better than its own mother can. At all events there is nothing in the treatment of hysterical children as important as placing them under the influence of a sober-minded, intelligent nurse or teacher who will devote herself or himself to studying the peculiarities of the child, and who will make a serious attempt to foster the good qualities and to counteract the vices. If this course that has been suggested is persisted in, it should be followed not for weeks or months but for years, and I am confident that if this is done during the formative period of a child's mind and character, an excellent result will follow in many instances. I base this upon my own experience, and have often stated to parents that if they will not allow me to pursue such a plan of treatment I prefer not to attempt any.

The Weir-Mitchell treatment, which is so effective in many cases of adult hysteria, is also of the greatest value in the hysteria of children; but I consider it to be one of its special advantages that it presents the first and easiest opportunity for the separation of the child from its immediate surroundings. Select a nurse carefully, place the child entirely in her charge, and many a parent will soon be convinced of the good that comes from such separation. In addition to the moral influences which may be exerted in this way the physical health of the child can be looked after. Bad habits of diet and of general hygiene can be corrected, and a child whose general physical condition has been far below par up to the time of its entering upon the rest cure, may be turned into a strong and vigorous being. Drugs, I am very certain, have but little influence over such conditions. The usual hysterical remedies, such as asafœtida, valerian, and the like, may be employed, but they will accomplish little which cannot be accomplished by other means as well. If the effect of a drug depends upon its disagreeable taste, suggestions of a different character will prove quite as efficient; and above all the physician should avoid the possible risk of engendering the idea in a growing child that there is a special drug which it may resort to for any annoying or painful sensation. There is nothing more disgusting than the habit so freely practised by many physicians of giving valerian or asafœtida or morphine to children or adults, whenever they present symptoms which smack of hysteria, or which cannot be interpreted otherwise by the attending medical adviser.

The special symptoms of hysteria call for distinct forms of treatment, but whatever these special symptoms may be, I always consider it wise to bear the general hysterical conditions in mind, and to employ, in addition to the special treatment, those general measures which have an excellent invigorating effect upon the nervous system. I refer more particularly to the proper use of hydrotherapeutic measures. The treatment of hysterical aphonia was referred to above. Simple faradism is all that is needed in most instances, and if this is not sufficient, regular vocal exercise in the hands of an intelligent teacher or nurse, will bring about the desired effect.

I have never found it necessary to apply the faradic current to the inside of a throat, but of course would not hesitate to do so in case the external application did not prove sufficient. Blistering or the application of the static current acts as a powerful form of local suggestion. In hysterical paralysis of the limbs, the use of a strong faradic current is generally the most effectual remedy. The current applied knowingly, not mercilessly, will gradually induce the patient to attempt similar contractions; and if to the use of the faradic current be added the encouraging words of the nurse or physician, who should not, however, accuse the child of simulation, and if other measures, such as massage, be employed, the hysterical paralysis will disappear in the course of time. But many of the cases are extremely stubborn and may require more vigorous measures, or more powerful suggestion, before they yield to treatment. It is of some importance at times to prove to the child that it can use its limbs very much better than it supposed. Place it in the middle of a room quite by itself, make it stand or crawl or walk, and if once shown that it can do so, the paralytic symptoms may rapidly disappear. But the treatment of hysterical patients of all ages and of all classes demands unusual tact and patience on the part of everyone concerned in the treatment of the case.

The sensory disturbances of hysteria are best influenced by the use of the cold douche or of the faradic current, particularly by the employment of the faradic brush. If such measures as I have suggested are not sufficient to remove an hysterical paralysis or an hysterical anæsthesia the effect of suggestion, hypnotic or otherwise, may be attempted.

The visceral disturbances common in hysteria also demand special treatment. Some of the measures to be employed were referred to in connection with the hysterical twins mentioned above. In cases of hysterical anorexia patient efforts should be made to induce the patient to take small quantities of food, and if these are not retained forced feeding must be resorted to; but never, if you can avoid it, resort to rectal feeding, for the patient who has discovered that she can be fed in that way will continue to refuse food

very much longer than she would otherwise. Lastly, the treatment of hysterical attacks may be managed in very much the same way as other hysterical manifestations are treated. First of all, the patient should receive the general anti-hysterical treatment, and should be given some drug which will act as a powerful irritant or a powerful form of suggestion when the attack is imminent. I have found nothing better than a sudden douche of cold water, or cold flagellations on the head and chest, or the inhalation of nitrite of amyl. The latter has this to recommend it, that it may be used with all the more assurance in those cases in which the suspicion of epilepsy cannot be altogether excluded. Lavender and ammonia are also efficient in some cases in which the hysterical attack can be inhibited by any such simple measure. If an hysterical attack continue for a prolonged period of time it may be brought to an end by the brisk use of cold douches, by the application of a strong faradic current, or by pressure over the ovaries, after the age of puberty, if these organs be oversensitive.

CHAPTER V.

CHOREA.

AMONG the neuroses of childhood none is worthier of careful study than chorea. The name is made by some to include a number of varying conditions, but its use should be restricted to designate a functional disease characterized by irregular, involuntary twitchings of some or all of the muscles of the body. These movements cease, as a rule, during sleep.

SYNONYMS.—This neurosis is also known as St. Vitus's Dance and the Chorea of Sydenham. Both these names deserve to be used, since the former implies its historical origin, and in the latter is preserved the name of the famous English physician who first described its most characteristic symptoms. Scelotyrbe and Melancholia saltans, are terms occasionally used, though almost obsolete. In German text-books the disease is called Chorea Minor, in contradistinction to Chorea Major, a neurosis of a purely hysterical character.*

ETIOLOGY.—Chorea is distinctly a neurosis of childhood and early adolescence. The vast majority of cases begin in very early youth, though Sinkler, many years ago, reported two cases in persons over eighty years of age. Careful statistics have been gathered with reference to this disease, the most elaborate being those reported by Dr. Stephen Mackenzie, in 1887, for the British Medical Association Collective Investigation Committee. Of four hundred and thirty-nine cases reported by this committee thirty-four per cent. occurred between the ages of five and ten years, forty-

* The name chorea, Greek χορεία, can be traced back to the dancing mania of the middle ages. During a severe outbreak of this psychic disturbance in Strasburg, in the early part of the fifteenth century, the chief magistrate of that city ordered those affected with this dancing mania to repair to the chapel of St. Vitus, in Zabern, a small village not far from Strasburg. The name, St. Vitus's Dance, is the only point of affinity between the dancing mania of old and the typical chorea of the present day.

three per cent. between the ages of ten and fifteen years, and sixteen per cent. between the ages of fifteen and twenty years. The largest number of attacks were found to occur in the thirteenth year of life. Some cases, however, occurred very much earlier than this average percentage would indicate. I have seen several cases of genuine chorea in children under one year of age, and many more in children up to the age of three years. That chorea is also occasionally congenital must be admitted: Sinkler refers to a case of this description due to fright of the mother during pregnancy. That there is hereditary predisposition to chorea is also evident, for the disease is developed more readily in children of choreic mothers and also in those whose parents are afflicted with epilepsy or migraine.

All authors are agreed as to the greater liability of the female sex. Within a period of three years I had seen 184 cases of chorea, in which 136 were females and 48 males. Sinkler among 328 cases reports 232 females and 96 males. Gowers, who has tabulated the largest number of cases, finds only 365 boys among 1,000 cases.

Dr. Weir Mitchell has studied the relation of races to the development of chorea, and claims that the negro race is almost exempt. I have no means of saying whether this is approximately true, but from my own experience I can assert that it is by no means rare among that race, but that among them it is if anything more frequent in boys than in girls. The disease is very common in Hebrews, as are many other neuroses.

Climate seems to exercise but very little influence upon the development of the disease proper, or upon the causation of the individual attack; it occurs quite as freely in cold countries as in warm, in northern as in southern latitudes, but there is an undoubted *seasonal* influence. The great majority of attacks occur in the spring. Drs. Mitchell and Lewis have made an elaborate research on this point, and claim that it is not so much variation in temperature or in humidity that causes the attacks, but that there is a decided correspondence between the number of attacks of chorea and the number of rainy and cloudy days; and then again between the attacks of chorea and the number of storm-

centres that pass over Philadelphia. Too much reliance should not be placed upon these statements until confirmed by similar researches in other cities. Putnam could not trace the same influences in Boston, and Gowers cannot accept the conclusions of Lewis for the conditions under which chorea occurs in England. These factors, if powerful at all, are of more importance in bringing about a recurrence of attacks that in giving the first impetus to the disease.

The causes which lead most directly to the development of St. Vitus's Dance are (in the order of their importance) fright, various acute diseases, such as articular rheumatism, scarlatina, and cardiac disease which so often accompanies the acute affections just mentioned. Gowers is inclined to regard fright as "the only immediate cause that can be traced with any frequency." It was the direct cause of chorea in 56 of my 184 cases. As a rule, the first symptoms of chorea appear within a few days of the fright; in some cases within a few hours, and even within the first hour. I had under observation for a long time a child that lived near the Brooklyn Theatre at the time it was destroyed by fire; the child was startled by the sight of the flames, and within a few hours began to twitch, and soon developed a severe attack of chorea which lasted for months. In one case the sight of a street brawl, in another the sight of a dead body, was directly the cause of the choreic attack. In children who have once had chorea very trivial occurrences are apt to bring about a recurrence. I have known a slight overstrain at school, the unexpected report of a pistol, a severe thunderstorm, or a scolding by a parent to be sufficient to bring on an attack.

The relation of acute rheumatism to the development of chorea has always been a matter of dispute. Some claim an absolute causal relation between the two, others insist that it is nothing more than a coincidence if one disease follows upon the other. Statistics upon this point are not so satisfactory as they might be, for cases have been reported in which it was stated that acute rheumatism has occurred, but it is not clearly made out that the rheumatism actually preceded the onset of the chorea. Sée reported the occur-

rence of acute rheumatism in about one-fourth of his cases, and in these figures he is supported by the conclusions of the British Medical Investigation Committee and by Gowers.* It is a curious fact the rheumatism does not seem to precede chorea nearly so frequently in earlier years as it does in those cases which occur between the ages of ten and fifteen years. From this we must infer that accidental coincidence plays a very much greater rôle than many are willing to concede. This must unquestionably be the case with the statistics furnished by Dr. Hamilton, who found twenty per cent. of all school children choreic or affected with some similar disorder. I could find a satisfactory history of the chorea coming on *after* rheumatism in only 20 of 184 cases.† Hirt, in his recent text-book on Nervous Diseases, thinks that there is a common toxic agent which, if it affects the cortex, will produce choreic movements; if it affects the joints chiefly, will give rise to acute rheumatism. This may be a rather hasty conclusion, but there is enough truth in it to say that the development of chorea is more probable in persons who have had rheumatism than in those who have not.

The majority of recent writers, including Herringham, Mackenzie, Oppenheim, Gowers, Osler, and others insist that an intimate relation exists between chorea and rheumatism; but the question arises whether heart disease, which is so frequently associated with rheumatism, may not be in part responsible for the development of chorea. Statistics, in order to be satisfactory, should be collected very much more carefully than they have been hitherto, in order to prove in how few, or in how many, cases of chorea the first attack has been preceded by rheumatism or cardiac disease. We can readily understand that fright should be a more powerful agent for evil in cases in which cardiac disease has preceded the existence of chorea. I have found satisfactory evidence‡ of cardiac disease preceding the development of chorea in only 20 of 184 cases. The bearing that

* Starr has tabulated 2,476 cases (by various authors); in 662 (twenty-six per cent.) there was a history of preceding rheumatism; in 502 there was cardiac disease.

† Osler states that there was a history of rheumatism in 15.8 per cent. of cases which he had examined carefully.

‡ The mere existence of a murmur has been taken by many as evidence of cardiac disease; in the fewest reports is there any accurate diagnosis of the cardiac condition.

heart disease may have upon the pathology of chorea we shall have occasion to refer to later on.

There is a marked tendency at the present time to overrate the infectious origin of chorea. One may claim that it is at times a post infectious disorder. To claim more would be unwarranted.

There has been much talk about reflex chorea, as about the reflex origin of many other neuroses, but he who sees with only half an eye will soon convince himself that these reflex theories are but a poor makeshift. Of all the cases of chorea that I have seen, I have found but very few that I could consider due to any peripheral exciting cause. I have convinced myself that in a few cases the presence of intestinal parasites was the cause of a transitory chorea, which disappeared as soon as the parasites were removed, but I am not convinced that nasal or ocular trouble, of which so much has been made of late, ever leads to true chorea. If these troubles prove an inconvenience to the child, some choreiform habits may for a time be established, but in such cases the cardinal symptoms of St. Vitus's Dance are wanting.

There is a curious relation between epilepsy and chorea. Gowers refers to epilepsy developing from chorea, and I have seen a case in a woman of forty, and another in a child of ten years, in which severe chorea set in after the cessation of epileptic attacks.

Symptoms.—Involuntary and irregular movements of any muscle or group of muscles of the body constitute the chief symptom of chorea. The muscles of the hands and fingers, and of the face and tongue are most often affected, but the leg and trunk muscles are at times involved. These movements are aggravated by volitional effort either of the muscles affected or of some other group of muscles. Thus the choreic movements of the hands will often become very much more intense if the child is told to stand absolutely still, or if while one hand is being examined it is asked to grasp something with the other hand. If the patient attempt to keep the affected part absolutely quiet he may succeed in doing so for a few seconds, but after that the movements will become more intense.

The choreic movements may affect only one extremity; they may involve one-half of the body (hemichorea), or they may be generalized. In 184 cases, 35 were cases of right hemichorea; 32 of left hemichorea; and in 117 cases the choreic movements were general.

As a rule the choreic movements are so evident that no special examination is needed. As the child enters the consulting-room the most superficial inspection is sufficient for a diagnosis; but in other cases, particularly during the early stages of the disease, the choreic movements are discovered only upon careful examination. If there is any doubt whatever about the condition, I ask the child to place its hand quietly upon my own, or between my two hands; the irregular choreic movements will, if present, be easily seen or felt. The true nature of the trouble, which may have appeared to be nothing more than "a slight nervousness," may thus be detected. If any further corroboration is needed, an examination of the tongue, as a rule, reveals the true character of the disease, for there are very few cases of chorea in which if the movements of the extremities are ever so slight, the tongue does not exhibit very marked choreic twitching. These tongue movements are slow, coarse, sometimes rhythmical. In advanced cases, if the tongue is protruded the mouth is opened much more widely than necessary, the eyelids and eyebrows are raised in the same effort, and then through a choreic movement of the masseters the tongue may be caught between the teeth. These movements combined give rise to what, in a former article, I called the "facies" of chorea.

The movements of the choreic patient are not only irregular but are often awkward to the extreme. This is clearly shown in the attempt to open or button the clothing, in raising a glass of water to the lips, or in attempting to hold the pen in writing. This awkwardness often induces great irritability on the part of the child; but however annoying the movements may be, it is only in a very small proportion of the cases that they lead to a condition of exhaustion. A few years ago I had occasion to observe a little girl, six years of age, the child of healthy and intelligent parents. It had passed successfully through one

attack of chorea, and in the second attack, coming on after a fright, the movements were extreme, and sleep was so poor that within a few weeks the child died from exhaustion. A weakened but not diseased heart unquestionably assisted in bringing about this early fatal termination. It is a fortunate circumstance that in almost all these cases the movements cease during sleep, and that the child is thus able to recover partially from the exhaustion caused by the movements during the day.

Some weakness of the muscles is frequently associated with choreic movements. The term paralytic chorea has been proposed for those cases in which there is marked paralysis, but as there is more or less weakness in the majority of the cases, and often more awkwardness than weakness, there does not seem to me to be sufficient excuse for the introduction of this term.

Speech is frequently involved. This is in the nature of a dysarthria rather than an aphasia, the choreic movements of the tongue and laryngeal muscles making speech difficult and often unintelligible. In some cases there is a little awkwardness of articulation, in others hasty articulation leading to the repetition of words, and in some a peculiar condition of speech which is in part due to difficulties of articulation, and in part to choreic movements of the respiratory muscles necessitating rapid breathing. Deglutition may be difficult, the tongue is frequently bitten, and from the awkwardness in the use of the knife and fork, and in passing food to the mouth, the little patient is much annoyed and is an ungainly sight for others while at his meals.

Laryngeal chorea, pure and simple, occurs somewhat rarely, and consists of choreiform movements of the muscles controlling the vocal cord. The result is a peculiar expiratory noise like a bark, which is repeated at short intervals. These cases are often mistaken for cases of hysterical bark; but the general restlessness, the age of the patient, the choreic movements of the tongue and fingers, should leave little doubt regarding the diagnosis. I remember a little girl, aged ten, who began to bark after a sudden fright; her case had been diagnosticated as hysteria

by several eminent physicians, but there was no element of true hysteria in the case. She recovered promptly under the usual rest treatment. There is little doubt in my own mind that this represents the rarest form of chorea.

The electrical reactions are sometimes slightly altered in cases of chorea. Rosenthal, Benedict, and others have found an increased response to the faradic and galvanic currents on the part of the muscles and nerves of the affected side. Some have even asserted that the reaction of degeneration with qualitative galvanic changes occurs in some instances, but I am inclined to doubt the correctness of this statement. I have never found a similar condition although I have frequently examined patients with this end in view. If such electrical changes were present I should suspect the presence of multiple neuritis, as this has been known to occur together with chorea; just as I might suspect this same complication in cases of marked sensory disturbances, for the rule is that in uncomplicated chorea sensation remains undisturbed.

Mental disturbance has been frequently referred to by many writers as a complication of chorea. It is surely not a very frequent occurrence, except that in the cases of chronic chorea (probably a different disease) the tendency to dementia is very marked. The impression I recorded a few years ago seems to me to represent the truth of the matter: "The mental calibre of children who develop chorea is rather above than below par. Children who by means of a better mental development stand head of the class, who work for prizes and earn them, children who are under constant mental strain, and about whom parents and teachers make much ado, are the ones most apt to be attacked by chorea." In some instances a violent mania is developed early (*chorea insaniens*), but it is much rarer to find this sequence of events than to observe cases of acute mania, particularly among young girls, in whom the movements of the extremities and of the tongue are typically choreic. Irritability of temper is perhaps the most frequent mental condition associated with chorea; but this is natural enough if we consider the very annoying movements and the difficulty the child sometimes expe-

riences in making itself understood. At times, instead of a condition of mania, a condition of apathy and depression is present in patients afflicted with chorea; but I am inclined to think that this is only true of patients who inherit a predisposition to mental disease.

The temperature has been studied in chorea. In mild cases it is normal throughout the entire course of the disease. In severe cases it may be raised a degree or two, but any greater elevation is undoubtedly due to some other condition.

Complications.—By far the most frequent complications are rheumatism and heart disease. Rheumatism, if present, is discovered easily enough both by the fever and by the painful swellings; but it is well to remember that the acute rheumatism of children is often a very much vaguer disorder than the acute rheumatism of the adult. If pain is much complained of in any case of chorea the joints should be examined carefully. Heart disease is the complication most to be feared. The heart should therefore be examined frequently and carefully. Mitral regurgitation is by far the most frequent form of cardiac disturbance. In the statistics of the British Investigation Committee there were 116 cases of mitral disease and only 6 of aortic disease. Gowers found but two instances of aortic regurgitation among 252 cases of chorea. Sinkler found cardiac murmur in 82 of 279 cases, but he does not decide how many were due to organic cardiac disease. It may often be difficult to determine this question, but if a patient whose heart was normal develops a murmur while under observation the probability of organic lesion is very great; yet since anæmia is very frequent in cases of chorea, we must allow, in judging cardiac conditions, for the possibility of hæmic murmurs and slight dilatation of the heart. Brown and J. K. Mitchell have described patients covered with subcutaneous nodules. These have a more direct relation to the rheumatic fever than to the chorea.*

An excess of urea and of phosphates has been found in the urine of choreic patients. It is questionable at best

* Osler is of the same opinion; he considers this condition a great rarity in this country.

whether they are not in some way the result of the incessant restless movements. Convulsive attacks are referred to as a complication by several authors. These are not of a typical epileptic character, but appear to be half-choreic and half-spasmodic movements. Mitchell and Burr have recently reported a case of this sort. The cases in which epilepsy is associated with chorea are more than likely cases of organic brain lesion, in which both the hemichorea and the epilepsy are symptoms of one and the same process in the cortex.

DURATION.—It is difficult to give any accurate information with regard to the duration of chorea, as the disease can hardly be said to be ended if upon the slightest provocation another attack sets in. A single attack may last from a few weeks to many months. The average duration is generally considered to be about ten weeks. In my own cases the duration of attacks varied between four and twelve weeks. Two and three attacks are much more common than a single attack. I found among 104 cases which were analyzed for this purpose that 50 cases had one attack; 17 cases had three attacks; 26 cases had two attacks; 7 cases had four attacks; 3 cases had five attacks, and 1 case had eight attacks. Notwithstanding this tendency to relapses the disease is an eminently curable one. It is only in a few cases that the disease becomes chronic, as in a patient of Meldner, who developed chorea in early life and remained choreic until his death, at the age of sixty-six years.

The interval between the relapses is also subject to great variation. In a few cases the relapse may set in after several weeks; in others after several years; and in the case of chorea of pregnancy we often find that ten years or more have elapsed since the preceding attack. The female sex, for reasons too evident to mention, is more prone to relapses than the male sex. The second and third attack is generally supposed to be milder than the first, but there are exceptions to this rule, for the very worst cases of chorea that I have ever seen have been in patients who were passing through second and third attacks. Later attacks, as a rule, simulate the earlier ones. If the first attack has been a hemichorea it is very probable that

later attacks will be of the same character. The severity of development, with the exceptions just mentioned, is very much as in the first or in the earlier attacks.

Diagnosis.—The diagnosis of chorea rests entirely upon the character of the movements. These are, as a rule, unmistakable, and are so typical that when they occur in connection with other diseases we speak of such movements as "choreic" or "choreiform." In practice the question is most frequently raised whether a child is suffering merely from general nervousness or from typical chorea. It has been my habit to decide this point not merely upon the character of the movements, though it would be safe enough to do this, but chiefly upon the presence of other symptoms, which I consider of still greater diagnostic importance. I refer particularly to the characteristic movements of the tongue, and to what I have previously alluded to as the "facies" of chorea. In rare instances a child may be able to imitate the choreic movements of others and thus simulate true chorea, but if it be mere simulation the attempt will not be a prolonged one nor will it be successful. Hysterical chorea can be distinguished very readily from the chorea of Sydenham by the more rhythmical character of the movements, by the peculiarity of the onset, by the longer free intervals between the attacks of twitching, by the longer duration of the disease, and by the presence of other stigmata of hysteria.

It is not generally appreciated that the choreiform movements associated with infantile cerebral palsies are apt to be mistaken for true chorea. This *post-hemiplegic chorea* is very similar to the ordinary form, but it is more strictly unilateral; it is more persistent, and it is invariably associated with other symptoms which prove the previous existence of paralysis. The difficulties of diagnosis are increased by the fact that in every case of severe chorea there is more or less weakness of the affected members, but in such cases I would advise examination for the existence of contractures and for increase of the reflexes, symptoms which are characteristic of preceding paralysis, even though there be little actual weakness at the time. Rigidity and increased reflexes, moreover, are never present in

cases of uncomplicated functional chorea. As this post-hemiplegic chorea is as much the expression of organic lesion of the brain as is post-hemiplegic epilepsy, it is natural that the choreic symptoms should continue as long as the cerebral lesion which has given rise to them continues in force. In a collection of cases of infantile cerebral palsies I have found this post-hemiplegic chorea present in about six per cent., from which it is evident that choreic movements are not nearly so frequently found in conjunction with these cerebral diseases as athetoid movements are. I have been consulted a number of times for persistent chorea, and in several such cases it has been my experience that the family physician has overlooked a preceding hemiplegic attack which could have been determined readily enough if attention had been paid to the existing contractures and to the increase of the reflexes. I would urge that in every case of chorea a careful examination be made for other evidences of organic brain trouble.

Confounding chorea and epileptiform convulsions is scarcely conceivable, for the convulsive movements of epilepsy come on at rarer intervals, there is generally some momentary loss of consciousness, and there are other symptoms pointing to epilepsy. In a previous publication I referred to the child of a colleague who would make sudden and very quick twitchings of an arm and of a leg. If these twitchings occurred while the child was walking or running across the room it would stand still, evidently surprised by these movements. It was natural to think of petit mal, but the frequency of the movements, the character of the twitchings, and the general choreic behavior of the child helped me to exclude petit mal and to recognize the case as one of true chorea. The diagnosis was corroborated by the very prompt result of antichoreic treatment.

Morbid Anatomy and Pathology.—In considering this part of the subject we meet with very much the same difficulties which we encountered with regard to epilepsy, and the resemblance between the two is also a close one in this respect, that we not only have a general functional disease, which in the one case we call epilepsy and in the other chorea; but, like epilepsy, chorea is also frequently enough

the expression of actual cerebral disease. It is natural therefore to infer that ordinary chorea must be due to disturbances similar to those which we find in cases of organic lesion. Almost every conceivable change in brain structure has been at one time or another held responsible for the development of chorea. Sée collected 84 cases of chorea on

FIG. 47.—Dilatation of Blood-vessels in the White Matter of the Convolutions of a very Chronic and Severe Case of Chorea. (Dana.)

which a post-mortem examination had been made. In 16 no changes were found in the central nervous system, in 32 there were lesions in the brain and in the nerve-centres; in the remainder there was congestion of the serous membranes. Ogle, Pye-Smith, and others refer to a hyperæmia of the brain and cord. As long ago as 1868 Steiner reported upon a careful examination of three cases of chorea. He found cerebro-spinal anæmia and some connective-tissue proliferation in the upper part of the spinal cord; conse-

quently he considered chorea to be the result of spinal irritation. Meynert and Elischer found hyaline degeneration in the nerve-cells of the central ganglia. The latter author also found changes in the vessels of the central ganglia as well as extravasation of blood into the connective tissue of the brain, and also numerous emboli in the smallest vessels of the cortex. He described peculiar corpuscles—highly refractile bodies—but Wollenberg has found them in the brains of non-choreic patients. Flechsig has found hyaline changes in the anterior divisions of the lenticular nucleus. More recently Dana has observed not only a general hyperæmia of the brain, but a degeneration in the walls of the blood-vessels, in the white substance of the brain, and considerable perivascular exudation with an accumulation of leucocytes. Many others, chief among them Hughlings Jackson, have insisted upon the embolic origin of chorea, a theory that would be plausible enough since Dickenson has found that in 17 of 22 fatal cases endocarditis was associated with the chorea; and yet this theory will not explain that large number of cases in which there is no involvement of the heart. Furthermore, an examination of the brain in fatal cases of chorea by competent observers has failed to reveal the presence of emboli. This view of the relation between capillary embolism of the brain and chorea was suggested by Angel Money, who noticed after injections of a fluid into the carotids of animals movements closely resembling those of chorea, and this condition was found after death to be associated with capillary embolism of the brain and cord.

Lockhart-Clarke found changes in the nerve-elements and connective tissue in the spinal cord; Garrod speaks of an overgrowth of connective tissue in the nerve centres, and thus we might go on quoting as many different findings as there are authors who have written upon this subject. One of the later contributions to this subject is by Anton, who found a lesion or old scar in the outermost division of the lenticular nucleus. As the same lesion was present in both halves of the brain, and the chorea was also a symmetrical one, the author is inclined to attribute the choreic disease to these lesions. By way of contrast

this same author reports the case of a man, sixty-five years of age, in whom spontaneous and associated movements were entirely wanting in the left half of the body. In the brain of this man the thalamus was very considerably diseased, and was supposed to be the cause of the defective movements. I need not discuss the author's theory attributing the excessive movements to the disease of the lenticular nucleus, and the defective movements to the thalamus. Other authors, basing their conclusions upon a number of autopsies, have attributed choreic movements to disease of the thalamus.

In an able article on rabies Golgi refers incidentally to his studies on cortical changes in chorea, which were published in 1874, and have been quoted since by v. Ziemssen and others. As a matter of historic interest the annexed figure is reproduced, showing the changes in the nerve-cells of the cerebellum; Golgi also reports that the ganglion cells of the cortex, and the cells of Purkinje, in the cerebellum, were calcified. It is doubtful, surely, whether there is any causal relation between these changes and others pointing to a chronic interstitial encephalitis, and the disease proper. Moreover, Golgi's patient died at the age of thirty-two years, and in him chorea was associated with chronic mental disease.

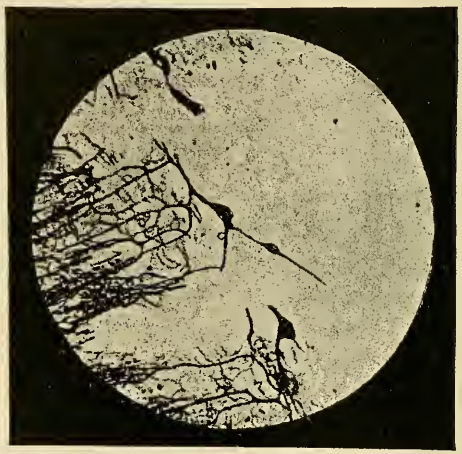


FIG. 48.—Changes in Purkinje's Cells in Chorea; Varicose Swelling of the Nerve-processes. (Golgi.)

Up to the present time the results of bacteriological research are not very promising. Berkeley found the staphylococcus pyogenes aureus in cultures from the blood of a fatal case of chorea.

Dana has published the history and autopsy of a case of chronic chorea. The patient was thirty-four years of age at time of death; he had his first attack of chorea at fourteen, and repeated attacks after that. The post-mortem findings included a chronic lepto-meningitis of the convexity of the brain, hyaline bodies in the brain cortex, slight meningitis of the upper part of spinal cord, and slight meningo-encephalitis. Diplococci were found in the proliferating tissue between the meninges and the brain.

The case is of unusual interest, showing that choreic symptoms may be

associated with a wide-spread affection; but the true pathology of chorea cannot be made out in any case of fourteen years' standing, nor can such a case be relied upon to prove the "germ-theory of chorea."*

The only just inference from the preceding account is that the accurate pathology and morbid anatomy of chorea are still unknown. Of the changes that have been reported by various writers, many, if not most of them, are secondary and not primary. All that we can claim at present is that there is considerable change in the gray matter of the central nervous system; that the entire motor tract may be involved, but that the changes occur more frequently in the cortex than in other parts of the brain. These choreiform movements are often associated with gross lesions in the cortex; they are for this reason more common in the child than in the adult, and a lesion anywhere in the brain so situated that it cuts off cortical impulses may give rise to chorea. The occasional development of mental symptoms, the association of chorea and epilepsy, the one following upon the cessation of the other, are the symptoms which not only indicate a cerebral but also a cortical origin. With more recent methods of examination, such as those described by Golgi and Cajal, we may be able to make out the permanent changes in the nerve-elements of the brains of choreic persons, but even such changes may be secondary to alteration of the blood-supply; in fact the tendency of the present day appears to be to regard chorea as due primarily to vascular changes, and such vascular changes may be the result of infection.

Prognosis.—Complete recovery is the rule in the majority of cases of chorea. The prognosis may be a little doubtful in regard to the recurrence of the disease and the duration of an attack. A child that has once had chorea has acquired a distinct predisposition to the disease, and often an occurrence which would leave no impression upon a healthy child's nervous system is sufficient to bring about a relapse of the disease. Later attacks are generally as

* Osler refers to the bacteriological researches of Pianese (Naples, 1893). Animals inoculated with a culture of bacillus taken from a choreic patient died "with muscular twitchings and convulsions," and the same bacillus could be obtained in pure cultures from the central nervous system. But it is doubtful whether these animals had chorea.

mild as the first, and there is no special reason to fear an unfavorable issue in later attacks unless severe complications set in. Under such circumstances not the chorea, but the complicating rheumatism, or endocarditis, is the actual source of danger. Death occurred in only two per cent. of the cases collected by the British Medical Investigation Committee, and Sinkler states that in Philadelphia, in seventy-four years, there have been but sixty-four deaths from chorea. This latter statement does not mean very much, as the majority of the cases of chorea ending fatally would be reported as cases of one of the complicating conditions.

There is no way of predicting positively the duration of a choreic attack. The milder an attack at the beginning the more likely it is to run a short course, whereas the severer forms are apt to be much more chronic, but severe cases under proper treatment will yield much more quickly than mild cases that are handled improperly. Under competent medical care the first attack may be recovered from in a period varying from four to ten weeks unless serious complications arise. A few get well more quickly, but they are the exceptions rather than the rule.

TREATMENT.—A severe case of chorea puts the skill of the attending physician to a severe test, while in a mild case the less medical interference the better for the child. Increased experience, both in private and in dispensary practice, prompts me to urge the simple plan of treatment which I outlined a few years ago.

The most important factor in the treatment of chorea is rest, absolute rest, often to the exclusion of all other therapeutic measures. Take a choreic child that has been accustomed to roam about at will and put it to bed; it will be a little restless for the first few days, but it will soon quiet down and show the great advantage of a thorough rest in bed. There is difficulty occasionally in carrying out this plan, for mothers and nurses are only too likely to be disheartened by the first show of resistance on the part of the child and its unwillingness to remain quiet; but with a few exceptions this morbid restlessness on the part of the child disappears within a few days, and the little patient feels very much

happier in bed than out. After a few days of enforced rest a decided improvement is noticeable. In the milder forms all movements cease, and in the severer forms the child is no longer troubled by the annoying jactations of the limbs. According to the severity of the case rest should mean entire rest in bed day and night. If the disease is taking a favorable course, after a week or two the patient may be taken out of bed for half an hour, an hour, or two hours, and then returned again for the remainder of the day. I have met only very few cases in which it seemed impossible to carry out this treatment; but I am firmly convinced that it was never the fault of the little patient but always the fault of incompetent and unintelligent relatives. If the disease has assumed a mild form we can endeavor to keep the child quiet without keeping it in bed the entire day; a few hours' rest will be better than none. It will also be of benefit to the child to forbid its taking any violent exercise, such as running, riding, dancing, or bicycling.

I am so convinced of the value of this rest treatment in chorea that I have made it a rule, even in dispensary practice, to insist upon this point of rest; we go to the extent of preferring the mothers to report to us about the child rather than to have the child taken out of bed and brought to us at short intervals.

Next in importance to rest is a nutritious and easily digestible diet. Milk and rest will do more for most cases of chorea than any other two measures. The nutritious diet will have a peculiarly good influence upon the many cases that are associated with profound anæmia.

The monotony of this special form of rest-cure can be varied with advantage by the use of lukewarm baths. Immersion into a cold bath, or the wet pack, with subsequent friction, cannot be recommended. It is better to place the child in lukewarm water, then reduce the temperature by adding cold water, and with this water, that is growing colder and colder, to sponge the spine thoroughly in order to get the effect of the dripping water upon the skin. After the bath the patient should be kept quiet and wrapped up warmly. In every case proper hygienic and dietary measures are of far more importance than medicinal treatment,

and yet we are bound to consider the various drugs that have been suggested for the cure of this disease.

Among these arsenic holds the first place. Its praises have been warmly sung by some, while others have decried it as but little better than any other drug that might be substituted for it. Only a few years ago Dr. Seguin, in his remarkably lucid lectures on the various forms of functional neuroses, placed arsenic first and rest second in the treatment of chorea. This order I think should be reversed, as I have yet to see the first case of chorea that got well more quickly with arsenic than without, as long as it was getting the benefit of rest. Dr. Seguin insists that physicians, almost without exception, give nearly useless doses of arsenic. He regards eighteen to twenty-seven drops of Fowler's solution after each meal as the really efficacious dose. In my own experience very few children will tolerate these large doses, which should at all times be given in some alkaline water within an hour after meals. I have always contented myself with smaller doses, varying from four to twelve drops three times a day, and if the cases resisted treatment I prefer abandoning arsenic rather than pushing it to the extreme which Seguin recommends. In cases of excessive restlessness I have been in the habit of prescribing the arsenic, together with the elixir of the bromide of sodium, or if the sleep is disturbed I give the evening dose only in this way and administer the arsenic alone during the day; the free use of chloral and bromides is to be condemned. If one must use any drug in the cases of chorea, arsenic is to be preferred; but it cannot be expected to perform miracles, and we must not regard it in any sense as a specific therapeutic agent.

Many other remedies have been proposed, all have been tried, and almost all have been abandoned. For some time the tincture of cimicifuga was in great favor. From fifteen to thirty drops, three times daily, may be administered in those cases in which arsenic is not well borne by the stomach. I have seen no good reason to resort to the use of hypodermic injections of arsenic, as recommended by Eulenburg and Hammond; Weir Mitchell some time ago proposed the use of salicylates; Simon and Legroux suggested the use

of antipyrin; and the oxides and sulphate of zinc have long been in popular favor in the treatment of this disease; but no one, I think, would venture conscientiously to recommend any of these drugs as a specific against chorea. The preparations of iron and of arsenic fulfil this *rôle* better than any other drugs, for they at least help to tone up the general system. Hirt recommends the use of morphine. Even if this drug is as effective as it is claimed to be, its use in young children is not to be encouraged. In the earlier stages of chorea it is essential for patients to obtain sleep. According to the age of the child chloral in five, ten, or fifteen-grain doses per rectum may be given. If it is necessary to substitute another drug I would suggest the use of chloralamid, of trional, of veronal (three to five grains at bedtime), and if there is a great deal of mental excitement I should favor the hypodermatic use of the hydrobromate of hyoscine (one two-hundredths to one one-hundredth grain).

In the majority of cases a heart tonic will be necessary. Digitalis in drop doses of the fluid extract or the tincture of strophanthus should be given in cases of heart weakness or feeble pulse. Blaud's pills, the sirup of the iodide of iron, various preparations of cod-liver oil, good stimulating wines, all these will be called for in some cases of chorea, but whatever else one may be induced to give the only matter of importance is that absolute rest shall be enforced.

Erb has advised the use of electricity. A weak galvanic current may be employed safely enough and may be applied to the nape of the neck, and over the motor areas; in which case, if given late at night, the current will help to bring on normal sleep. A moderate stabile current of from fifteen to twenty cells (about ten milliampères) applied to the spine will help to allay restless movements of the body. Massage may be given in some cases in which the general nutrition is at a very low ebb and in which the circulation is poor.

A special caution is necessary as regards the question of attendance at school. Every choreic child, however mild its attack may be, should be kept from school both for its own sake and for the sake of the other pupils who might imitate the disease. I have sometimes allowed myself to

be persuaded to permit a child with a mild form of chorea to continue at school; in almost every instance I have had reason to regret it, for nothing is better calculated to bring out severe chorea than the competitive spirit that obtains in most schools. Periods of examination are fraught with greatest danger to those children who have had attacks in earlier life. The atmosphere of the school-room seems to have a depressing influence upon such children, and among the wealthier classes far better progress can be made in the ordinary studies if the child is instructed at home than if it is taught at school. It is necessary for the physician to take a firm stand on this question or else his treatment of the case will be thoroughly unsatisfactory.

CHAPTER VI.

CHOREIFORM DISEASES.

HEREDITARY OR HUNTINGTON'S CHOREA.

THIS disease is, on the whole, extremely rare. In view of its hereditary character we must consider it in connection with the diseases of children, although it generally appears about the age of thirty or forty. The disease was first observed by Huntington, a Long Island physician, and it has since been designated by his name. It appears, however, that another New York physician, Waters, described the disease in a letter to Dunglison. For a long time the disease was little known beyond America. Of later years it has been observed and discussed by a number of English, German, and French authors. The disease runs in families and is spread from one community to another in connection with the migration of the afflicted families, so that now, according to Gray, there are a number of communities in America in which the disease is prevalent; but, as he further states, there is great secrecy maintained with regard to it, as the affliction is looked upon as a distinct stigma resting upon the family.

The disease affects several members of the same generation, but may skip as many as it selects. The descendants of healthy members of a family enjoy immunity from the disease as a rule. In King's case, which I quote on the authority of Gray, the great-grandfather was choreic. He had ten children, but only four were afflicted with this disease; they also had children who were choreic. One of these four had nine children, eight of whom were healthy, but the ninth was choreic. The ninth had five children, of whom four were choreic. Of these four, three had no children, but the fourth had a chorea whilst he was still young and

was cured of it, when again at thirty-five he passed gradually into the typical Huntington's chorea.

Symptoms.—The disease begins, as a rule, between the ages of thirty and forty years, though at least a dozen or more cases are on record which have begun before the age of twenty. Males and females seem to be afflicted with the disease in equal numbers. The chief symptom is a motor disturbance, which for lack of a better name is termed choreic, yet it is very different from the twitchings of chorea minor. The muscular movements of Huntington's disease are coarse and grimacing, and may be distributed over a large area of the body. In Sydenham's chorea the affection is more strictly localized. In the former disease grimaces and all sorts of extravagant posturing are much more pronounced than those which we observe in the ordinary chorea of children. The difference in degree of muscular activity is so very great that one naturally doubts the connection between the two diseases. As a rule, the movements are slight at the start, affecting the face and upper extremities only. In the course of years they increase in intensity, and become widely distributed over the entire body, until in the end every single muscle or group of muscles may be involved.

The muscular disturbance is in part subject to the will, and patients afflicted with this disease can, if they make a serious effort, inhibit the twitchings very much better, and for a much longer period, that those suffering from ordinary chorea can. The motor power is, as a rule, not diminished, but on account of the irregularity of muscular actions it is extremely uncomfortable for the patient to walk about, and he easily becomes an object of pity or of ridicule.

There are no sensory disturbances, and the reflexes, while they may vary much, are not morbidly exaggerated or diminished. By far the most characteristic symptom of Huntington's chorea is the association with it of a progressive dementia. The first sign of a mental change may be a simple depression which is deepened by a knowledge of the hereditary and incurable character of the disease. Under the circumstances the occurrence of suicide is not unnatural. The mental condition at the start varies a little ;

the patient may be either irritable or apathetic. In the course of time the deterioration of all the mental faculties is very marked and a typical dementia is developed. Speech is, as a rule, thick and indistinct, sometimes nasal and confused. Unless some intercurrent disease puts an end to life these patients may linger on for many years, may become absolutely bedridden, a burden to themselves and a torment to their families.

The course of the disease is a very chronic one, and unfortunately does not tend to shorten human life. Several of those who have exhibited the first symptoms of disease at the age of thirty have lived to sixty and seventy years. Whether cases beginning very early in life live as long, cannot be distinctly stated, but there is nothing improbable in supposing that they do.

ETIOLOGY.—Among the causes of this disease none is more potent than heredity. It is distinctly a family disease, and unlike other such diseases it does not seem to skip a generation. Persons with this disease can generate healthy children whose descendants may not be afflicted with this trouble. To all appearances an exciting cause is needed to develop the disease. In many of the cases the first symptoms appear upon severe emotional excitement or after some acute disease. In this respect the resemblance between the hereditary chorea and chorea minor is very striking. Rheumatism does not, however, play the part in the hereditary form that it does in ordinary chorea.

DIFFERENTIAL DIAGNOSIS.—The strong factor of heredity and the appearance at a relatively advanced age are sufficient to distinguish these cases from ordinary chorea. There is some danger, however, of confusing these cases with the post-hemiplegic chorea that is developed early in life, and this danger is all the greater if the onset of the paralytic symptoms is uncertain, or if the paralysis has disappeared and the chorea remains. From Friedreich's ataxia the disease can be easily differentiated by the fact that in the former malady the disturbance of motion is truly ataxic and not choreic, and the reflexes are totally absent, whereas in Huntington's disease the reflexes are not markedly altered. In Friedreich's disease there is also

an ataxic gait, made worse by closing of the eyes, whereas in Huntington's disease nothing of the sort is observed. Some forms of hysterical chorea might be confounded with this disease, particularly if on inquiry the statement is elicited that a similar affliction has been observed in other members of the family, and it should not be forgotten that the hereditary trouble may be of an hysterical order. The examination of the patient and the general hysterical temperament, the fact that the choreic movements come on in the nature of attacks, and the determination of other hysterical symptoms will help to distinguish one disease from the other.

Pathological Anatomy.— The morbid changes in Huntington's chorea have not yet been definitely made out. Innumerable changes have been reported by various authors. These include pachymeningitis, hæmatoma, tumors of the dura, general atheroma, atrophy of the motor convolutions, increased fluid in the ventricles, foci of softening in various parts, even in the ganglia. The most frequent states are pachymeningitis and changes in the cortical tissue. In a case of Charcot's the meninges were adherent in certain places, and the cortical substance was evidently sclerotic. But these varying conditions are largely secondary processes and do not in any way explain the true pathology of the disease. Dr. Osler summarizes the changes in a series of sections from the brain and cord which he examined as follows: "The arteries were thickened, and, in places, show hyaline degeneration, and in the smaller arterioles the fatty changes were very marked in the fresh specimens from the cortex. The perivascular lymph-spaces were large, and contained leucocytes. The ganglion cells, in many sections, showed very slight changes, not more than are often seen in chronic cases associated with atrophy of the convolutions. There was the common vacuolation, and many cells seemed laden with pigment. The increase in the connective-tissue elements was more evident to the touch and upon section than microscopically. Sections of the pons and medulla showed no foci of disease. Beyond the thickening of the arteries and the shrinkage of the cells of the anterior cornua—probably an artificial

change—the sections of the cord showed no important lesions."* Oppenheim, Hoppe and others found disseminated, miliary encephalitis of the cortex. Binswanger thinks the morbid changes not unlike those of general paresis. French authors attach great importance to the proliferation of the neuroglia of the cortex.

PROGNOSIS.—The prognosis of Huntington's chorea is grave as regards the cure of the trouble, and in view of the marked dementia which is associated with almost every case. Unfortunately, however, it is not a fatal disease, and patients so afflicted may live on to a very old age.

TREATMENT.—As for the treatment of these cases the same principle should be observed as in ordinary chorea—rest, freedom from care, and excitement, separation from family, and change of climate, and possibly a course of arsenic treatment may be of some benefit. If movements are excessive and the sleep of the patient is unsatisfactory, the exhibition of hyoscine, in doses of one one-hundredth of a grain, or of trional or chloralamid, in ten- to twenty-grain doses, once or twice a day, is worthy of a trial. But, in the nature of things, every form of treatment will be simply palliative.

HEREDITARY CHOREA WITHOUT DEMENTIA.

There is another form of chorea of which a slight mention should be made in this connection. The disease has a distinct hereditary tendency, and is either transmitted direct from parent to child or from a more remote ancestor. The disease appears early, beginning as a rule at about the age of puberty and continuing during life. It is characterized by distinct choreic movements of the hands and tongue and the facial muscles. The movements may become so marked that they interfere with every voluntary effort, such as riding, sewing, buttoning of clothes, and the like. The lower extremities do not become affected, and the general health of the patient is not much impaired, except that the annoyance of the disease may bring on a despondent feeling; and in one case that has come under my notice distinct melancholy has occurred. It is distinguished from Huntington's chorea above all by the entire absence of symptoms pointing to dementia. The persons afflicted with this form remain bright throughout life and are able to attend to their ordinary affairs. It is furthermore to be distinguished from chronic chorea from the fact that there is no distinct history of an acute attack after which the chorea has been developed; that the choreic move-

* Greppin claims that the disease is due to a morbid process (not unlike an encephalitis), which is developed gradually and in various parts of the brain.

ments have never been as wide-spread as they are in such cases of chorea minor as develop into chronic chorea. They generally begin a little later in life, too, than the acute chorea minor does, and there is no distinct history of individual attacks. If the disease has once been established it continues without any marked remission, but also without any rapidly progressive changes. Some confusion might also arise with the post-paralytic chorea, but under such circumstances an examination into the past history of the patient, and for the evidences of persistent paralysis, will help to clear up the diagnosis.

The prognosis is entirely favorable as regards life, but less can be said as regards the cure of the disease. I have placed such patients under the complete rest-cure, and have for a time obtained distinct improvement in the choreic movements. General tonic measures should be employed, and the proper feeding of the patients carefully looked after.

HABIT CHOREA.

It is well known that children are subject to "tricks" of movements. These include simple or co-ordinated movements of various muscles, generally of the muscles of the face, of the eyes, of the shoulder, and even of the thighs. These "habit spasms" or "tics" may resemble chorea; often the jerkings are so violent in character as to suggest "epileptoid" rather than "choreic" disease.

It is not always an easy matter to determine how the "trick" was engendered; in some instances a true chorea has been the starting-point, the children keeping up some form of twitching movement after the St. Vitus's dance has disappeared. Through imitation, habit chorea may be developed in children who have watched others with chorea minor.

A habit chorea developed in early years is often continued throughout life. Among professional men (artists, literary men, and even physicians) such "tricks" are not uncommon, and the doubt arises whether serious effort has been made to dispel the habit. Among the commoner forms of "habit chorea" are blinking, facial contortions, sniffing, shrugging of the shoulders, or some trick in speech or gesture. A friend of mine, now a well-known astronomer, has since boyhood never answered a question without first saying, Hm? ah? eh? In former years this utterance was accompanied by an exaggerated raising of the eyebrows, as if to intensify the interrogation.

Such habits cannot be condemned sufficiently in children. Parents should discipline children severely in order to rid them of the habit.

If there is any peripheral condition (such as nasal obstruction or eye-strain) which helps to keep up the habit, the condition should be treated carefully. It is important also to make certain that the "habit spasm" is not true chorea. This can be done by examining for other symptoms of St. Vitus's dance. The general condition of children with some form of habit spasm may require treatment; many of them are anæmic or scrofulous.

Complex Co-ordinated Movements (Complex Tics).—In children as well as in adults, complex movements of a definite character are repeated

at intervals, or may be continuous. They bear only a very slight resemblance to chorea; but, as a matter of convenience, they may be discussed in connection with "habit chorea." Some of them, the gyrospasms, for instance, occur in very young infants; others occur in older children and are frequently associated with slight mental derangement. Some authors have included them under the heading of Imperative Movements, and have described them as head-shaking, head-rotating, head-banging, head-nodding, etc. These movements are often associated with nystagmus and with defective mental development, thus indicating that in some instances they may be due to organic changes in the brain. These peculiar disorders have been studied carefully by Henoch, Hadden, Gee, Peterson, and Mills, and have been referred to by Osler and others.

Gyrospasms of the Head is a term applied by Peterson to peculiar rotary movements of the head in children, associated at times with strabismus or nystagmus. The movements of the eye are very rapid (Risien Russell) and may affect one eye only; the nystagmus may be of the convergent order (J. Thomson). Two of Peterson's five cases were observed by him in my clinic. Both these cases were in young infants, eight and nine months of age respectively; in one the movements came on after a fall; in the other there was no history of traumatism. The chief points of Case II., as described by Peterson, are as follows: The child was nine months old when examined. Since the age of four months he had a rotary movement of the head. When the child was quiet, the head kept oscillating from right to left and from left to right at the rate of eighty to one hundred oscillations per minute. The movement ceased at times, particularly if the child's gaze was riveted upon some object. There was lateral nystagmus of the right eye, also ceasing for minutes at a time.

These gyrospasms, which are evidently of a piece with head-nodding, head-jerking, etc., are scarcely to be confounded with *epilepsia nutans*. If developed in later years, they may constitute a habit, but if the movements begin in early infancy and are associated with nystagmus, strabismus, or idiocy, one need not hesitate to ascribe them to cerebral disease. The region of the cranial-nerve nuclei would be the most probable seat of the trouble; and the lesion must be supposed to be irritating in character.

The chief etiological factors appear to be rickets, intestinal irritation, and dentition. The prognosis is good on the whole, except when these movements are associated with idiocy. A mild course of bromides has proved satisfactory in the majority of cases. Mills recommends, in addition, the use of two or three minims of tincture of belladonna, or one minim of the fluid extract of conium.

Imperative movements associated with arithmomania (repeating everything a definite number of times) are clearly the result of mental derangement, and do not seem to me properly to belong to this group of cases.

Several other motor neuroses bearing a more or less close resemblance to chorea minor have been described in

connection with the chief disease. While the cases may have a superficial resemblance to St. Vitus's dance they are, as a rule, of a much more explosive character. They occur in families with a neurotic heredity, and represent, on the whole, a more serious disturbance of the central nervous system. It is not an easy task properly to classify all these conditions. There has been much confusion in the discussion of these subjects, and no two authors entirely agree in the designation of these disorders. As a majority of them come on later in life, or at least are fully developed in later years, we need not in this book treat them in a very exhaustive manner.

CHOREA ELECTRICA.

It is certain that two distinct forms have been described under this term. Henoch includes under it a form of choreic disturbance which resembles in part what is commonly designated as habit spasm, but some of his cases are more distinctly allied to myoclonia or paramyoclonia multiplex. Bergeron alludes to violent choreic movements occurring chiefly in children between the ages of seven and fourteen years. Bruns considers these two forms to be identical. Oppenheim is inclined to label Bergeron's form as an hysterical chorea.

The term electric chorea should, possibly, be restricted entirely to a very rare disease that occurs chiefly in the northern part of Italy. I have had opportunity to see one case of this description in Italy, and one in an Italian woman who came to my clinic. The condition was first described by Dubini, in 1846, and to avoid confusion it would be as well to speak of it as Dubini's disease. The disease is apt to occur in boys and girls as well as in men and women of advanced years. It has been supposed to be due to some infectious agent, but the rarity of the disease, even in those districts in which it does occur, would seem to militate against this view.

The chief symptom of the disease is a series of violent spasmodic movements, affecting particularly the muscles of the neck and head, as well as of the extremities. Some of the cases begin with movements in an arm, which spread to the leg of the same side, and finally involve the opposite half of the body, and also the trunk, neck, and head. After some months the choreic members of the body become paralyzed; there is wasting of the muscles and a loss of faradic irritability. Epileptiform seizures occur, and these may either be partial or general. In the course of a year or more the patient may become entirely paralyzed, and, as in the case I saw, the patient was confined to his bed, unable to move a limb, while the severest choreic jactation continued in the muscles of the neck. The majority of these cases end fatally within a few weeks or months. During the course of the disease the patient suffers a great deal from pain, has slight elevation of temperature, but, as a rule, does not have loss of consciousness.

The disease is so rare that sufficient pathological examinations have not been made to warrant definite statements with regard to its nature. It is probable that it is a form of cerebro-spinal disease, as is indicated by the convulsions on the one hand and the paralysis with atrophy on the other.

MALADIE DES TICS CONVULSIFS.

This disease, also known as Gilles de la Tourette's disease, has been studied most carefully by Gilles de la Tourette, Guinon, Brissaud, Meige, and Feindel (who have written a monograph on the subject), Oppenheim and others. The chief symptoms are convulsive twitchings of the facial muscles and other regular systematic movements, explosive conditions of speech known as echolalia and coprolalia, and lastly, imperative conceptions and impulses. The disease, as a rule, appears in children between the ages of seven and fifteen years. It is more apt to occur in those predisposed by inheritance to neurotic troubles than in others. In many instances there is a very distinct history of heredity, the same disease occurring in succeeding generations. (Charcot, Gintrac, etc.)

The first symptoms that appear are sudden and explosive twitchings of the muscles of the face or of the eyes. The mouth may be twisted, and all sorts of grimaces may constitute the early symptoms. In a case which I have had under observation, the boy would begin by making grimaces and then turn about quickly as though he was snapping at some one. These movements were performed in regular succession. Movements of the sterno-cleido mastoid, and of the trapezius are often repeated so systematically that they seem to be purposive. The child may at the same time begin to spit or to blubber, and the entire combination of symptoms suggest the possibility of simulation or intention, particularly if jumping or leaping movements are associated with the other symptoms. Whatever the symptoms may be that have been developed in a given case they are persisted in with remarkable regularity, and can be distinguished from intentional movements by the fact that the patient is evidently surprised by the suddenness and violence of the movements. Smacking, hissing sounds are sometimes heard, but none of the symptoms is more characteristic than the habit of repeating words or sounds that are heard (echolalia), or the involuntary sudden explosive utterance of foul language (coprolalia). A little patient of mine uttered the worst curses I ever heard, which she had evidently picked up on the street, but would regret the utterance the very next moment after they had passed her lips, and would contritely declare that she was entirely irresponsible for the same.

The symptoms frequently continue for years, and while the intelligence of the person so afflicted does not show any deficiency only a few are able to become masters over these explosive seizures. In some an attempt to conquer the disease produces great restlessness and general excitement. In others the symptoms become manifestly worse under the effort to control them, and it is advisable under such conditions to divert the attention of the patient from his symptoms as much as possible.

Imperative conceptions are frequently associated with the motor symp-

toms, and usually seem to represent a psychic explosion, the equivalent of the physical symptoms. Some of the children feel compelled to utter words in a definite sequence, to pronounce certain letters in a peculiar way; rolling the "r" and making a hissing "s" are particularly frequent. In others imperative actions, such as occur in connection with neurasthenic disorders, are quite common. They are compelled to do their sums over and over again, to retrace their steps, to pick up everything they see lying on the street or in the room, and are compelled to be in a state of continued activity in consequence of these imperative impulses.

The course of the disease is extremely chronic; by some it is considered to be absolutely incurable. But this is too extreme a view, since some of the cases get well.

The prognosis should, however, be studied carefully in each case, as there is no telling in advance whether the patient will respond favorably or not to the treatment. The resemblance between this disease and other choreiform neuroses is at times so close a one that a differential diagnosis becomes extremely difficult. It can hardly be confounded with ordinary chorea since the movements are much more systematic and more intermittent than they are in St. Vitus's dance. Moreover, echolalia and coprolalia never occur in ordinary chorea. The difficulties of speech in chorea are the very reverse of the explosive speech in this maladie des tics. Hysteria and hysterical chorea may bear a superficial resemblance to this disease; but, on the one hand, the stigmata of hysteria are generally wanting, and in hysteria there is no such constant repetition of more or less convulsive movements as there is in this disease. There is, no doubt, a close resemblance between the maladie des tics and the jumpers and many other forms of disease which are differently designated according to the peculiar variety of muscular action. The one point, however, which the maladie des tics has in common with other similar conditions is its occurrence in neurotic families. The distinction between maladie des tics and para-myoclonia will be evident after a discussion of the latter trouble.

TREATMENT.—The most important factor in the treatment of these patients is their complete isolation from all other children and from their usual surroundings. They should be subjected to a moderately severe discipline by a competent nurse or parent, and should be taught self-control as far as that may be possible. The mere seclusion from other persons generally serves to lessen the excitement and the number of explosions. If the disease has led to a condition of exhaustion from loss of sleep or from inanition it is well to secure quiet during a considerable part of the entire day. This can be done best by the administration of chloral or of small doses of morphine. I cannot see the wisdom of using chloroform, as has been suggested by some. If the motor restlessness is extreme, hypodermic injections of hydrobromate of hyoscine, in doses of one one-hundredth grain, carefully administered, may prove of some benefit; the ordinary nerve-tonics, such as arsenic and quinine, will do little good, and it is best not to waste much time in administering these drugs. I am confident that if any good is to be accomplished we must depend entirely upon isolation, proper feeding, and the use of tonic hydro-

therapeutic procedures. Regular gymnastic or calisthenic exercises should be tried and may be of advantage.

THOMSEN'S DISEASE (MYOTONIA CONGENITA).

In connection with the various forms of disordered movements which have been considered in this chapter, we may discuss in a very brief way the disease known by the name of the physician who gave the first thorough description of it as it occurred in his own family. Those who object to nomenclature of this sort will prefer the term Myotonia congenita, but as Thomsen was also a subject of the disease an exception may well be made. Very few cases have been described in America, and the only one which the author has had an opportunity of seeing was the one presented to the New York Neurological Society, in 1886, by G. W. Jacoby. The best accounts of the disease are those furnished by Thomsen himself, a description of it in Leyden's work on "Spinal Cord Diseases," and the monograph of Erb, who summarized the chief points of twenty-two cases, and added a full account of peculiar electrical changes in this disease, for which he proposed the term "Myotonic Reaction." Dejerine and Sottas, Schiefferdecker and others have more recently contributed to the pathology of the disease.

The chief symptoms of the disease are a rigidity of the muscles (myotonic contraction), which is developed whenever an attempt is made to use a muscle or muscles after a period of rest. If the first difficulty has been overcome, the action of the muscles may be entirely normal for some period of time. This muscular rigidity is most noticeable in the attempt to rise after a person has been quietly seated for some time; in the attempt to use the hands in lifting, grasping, or writing, and at times considerable difficulty is experienced in the movements of the tongue and of all muscles concerned in articulation. The impression created by a patient grasping the physician's hand and then not being able to let go, or by his falling to the ground and remaining absolutely rigid until the spasm is relaxed, is one not easily forgotten. The spasms are generally limited to a few groups of muscles, but in some cases the entire muscular system, with the possible exception of the ocular and respiratory muscles, is involved. The rigidity of the muscles is increased under the influence of emotional excitement and under the influence of cold and damp weather. Under alcoholic stimulants the rigidity is said to lessen, and prolonged periods of rest undoubtedly have a favorable influence upon the disease.

The disease occurs in families, and very often affects several members of the same family. In Thomsen's family, five generations have been affected by this same condition. There seems to be no distinction as regards sex, and the disease may come on at any period of life. A number have been recorded during the first decade of life, but the disease is apt to reach its maximum during the period of greatest muscular development, and the symptoms are, therefore, pronounced between the ages of fifteen and twenty-five. In Jacoby's case the patient, who was twenty-four years of age at the time of

examination, stated distinctly that he could not, as a boy, take part in the outdoor games of his comrades on account of the stiffness of his muscles. He attempted to play upon the organ, but found that his hands were clumsy. He was not able to whistle, and even in chewing his food the muscles became stiff and rigid, simulating a condition of trismus. In this case there was distinct rigidity of the muscles of the eyes.

The entire muscular system is, as a rule, well developed; there is often an increase of normal volume. According to Thomsen, the more the muscles are used the less the spasm becomes, and he advises a very active life as possibly the only hope for an improvement in the disease.

There are a few objective symptoms which render the diagnosis of the disease easy enough. The mechanical excitability of the muscles is markedly increased, a single tap of the hammer on a muscle produces a slow, tonic contraction of the fibres, but the contraction is not relaxed for some period of time. In contradistinction to the symptoms of tetany it may be stated that the nerves do not show any increased mechanical excitability. There is, furthermore, a very remarkable change in the behavior of the muscles under electrical stimulation. Erb has proposed to speak of this as "myotonic reaction." The faradic excitability of the nerves is not changed, but on the use of a strong current the muscles innervated by the nerve will be forcibly contracted and remain contracted for some period of time after the current is broken. The direct faradic excitability of the muscles is very much increased, mild currents being sufficient to produce contractions of long duration. The galvanic excitability of the nerves is surely not increased—possibly diminished—but the same muscular phenomenon can be observed on galvanic stimulation of the nerve as in the case of faradic excitation. The direct galvanic excitability of the muscles is increased; the anodal contraction is, as a rule, greater than the kathodal contraction; opening contractions either with the anode or kathode cannot be readily obtained. The contractions are sluggish, and are continued for some time after the electrical stimulation ceases. But the most characteristic symptom of this myotonic reaction is a peculiar, rhythmical, wave-like contraction which proceeds from the kathode toward the anode. This phenomenon can best be observed with the use of strong currents of at least 20 or 25 milliampères, and if the negative pole is placed over the tendinous end of a muscle. If the kathode is placed in the palm of the hand, the anode on the shoulder, a wave-like contraction will appear and gradually work its way from the muscles near the wrist to those of the shoulder. The response to the static current remains normal.

These are the chief symptoms of the disease. A few complications have been observed in some of the cases, among which we may specially mention increased mental irritability and hypochondriasis, both of which are not unnatural in view of the annoyance which the disease causes the sufferer. Epilepsy and migraine have also been recorded in connection with Thomsen's disease, but as both these diseases are to a great degree hereditary the association may be simply a coincidence.

The pathology of the disease is still obscure. Dejerine and Sottas found

no changes in the nervous system, but endorse the findings previously made out in muscular tissue excised from the living body. The changes noted were an enormous hypertrophy of the fibres, with considerable increase of the nuclei in the sarcolemma, and an increase of the interstitial connective tissue. In one case examined by Erb, there was a striking vacuolization of the individual muscular fibre. These histological findings do not enable us as yet to explain the character of the disease. Thomsen considers defective cortical innervation to be the prime cause, but cerebral changes alone could not account for all the symptoms; these must be ascribed to disease (primary or secondary) of the muscles or to disease of the cord. Some have been inclined to attribute the disease to auto-intoxication. Proof is wanting; the findings point merely to an abnormal development of the muscular system.

The disease can be mistaken only for tetany, and possibly for chronic muscular dystrophies; but as Erb suggested, several years ago, a tap with the percussion-hammer and a few closure contractions with kathode and anode upon certain muscles will suffice for a diagnosis of Thomsen's disease.

The prognosis is favorable as regards life, unfavorable in regard to a cure of the disease. Much can unquestionably be done by a continued exercise of the will and by regular gymnastic exercise. No further treatment of the disease is indicated.

CONGENITAL PARAMYOTONIA.

Eulenburg * has described a somewhat similar affection, which he terms "Congenital Paramyotonia." This also is a distinctly family disease which may be traced through six generations, the disease becoming manifest immediately after birth. The chief feature of these cases is a tonic spasm lasting from a quarter of an hour to several hours, and coming on chiefly after exposure to cold. There is distinct weakness associated with rigidity; the facial muscles may be involved as well as the orbicularis palpebrarum and the orbicularis oris, making speech impossible during the continuance of the contraction. The disease is differentiated from Thomsen's disease by the absence of increased mechanical excitability and by the absence of anything resembling a myotonic reaction. Eulenburg states that there is a distinct tendency to kathodal and anodal tetanus.

PARAMYOCLONUS MULTIPLEX (MYOCLONIA).

A short reference is necessary in this connection to the above disease, which bears a resemblance to electrical chorea and to tic convulsif. By some it has been regarded as a form of hysteria, but this is not warranted, for cases have been observed in which there were none of the characteristic signs of hysteria. It is best to restrict the term paramyoclonus multiplex to the disease as it was originally described by Friedreich. The chief symp-

* Neurologisches Centralblatt, June 15, 1886.

tom of the condition is a rapidly repeated clonic spasm, occurring in attacks and affecting individual muscles or groups or muscles, and as a rule symmetrical muscles are involved. The muscles of the extremities are more frequently affected than those of the other parts; the face muscles are generally exempt, and in this respect the disease differs markedly from other forms of choreic and clonic spasm. These clonic convulsions are not severe enough to cause actual movements of the extremities, but in a few cases sudden contractions of the diaphragm have been accompanied by hiccough and by peculiar respiratory sounds, and in the case described by Starr the movements were strong enough to produce jactitation of the head and of the trunk. In contrast to some of the other diseases described in this chapter, the myotatic irritability is increased only a little or not at all. Electrical excitability is not altered.

The disease may come on at any period of life between the age of puberty and fifty years. It is rare in young children, and the short account of it here is simply given for the purpose of showing its close relationship to the various forms of choreic disorders. Heredity is not a prominent factor in the cases which have been described, but it has been found in several members of one family. Mental or physical strain and emotional excitement are the chief causes. Cases closely resembling, yet not identical with paramyoclonus multiplex have been described by Hammond and others, under the title of "Convulsive Tremor."

The prognosis is serious as regards a cure of the condition, though some of the cases have been reported improved and a few entirely cured. In the treatment of the disease nothing more can be attempted than regular gymnastic exercise, and the exhibition of mild sedatives, such as chloral and the bromides, in small doses, in case the spasms are excessive. A mild galvanic current is said by some to have exercised a favorable influence over the disease.

Dana has endeavored to establish a classification of myoclonias approved by Lundborg. It is as follows: 1. Myoclonia of Friedreich, or peripheral type including myokymia; 2. Myoclonia of the functional or hysterical type; 3. Myoclonia of the convulsive tic type (many associated spasms, choreic and tonic); 4. Myoclonia of the "familial" type or myoclonus epilepsy—closely associated with the third; 5. Myoclonia of the type of the infectious and symptomatic choreas. (Clark and Prout.)

MYOCLONUS EPILEPSY.

Unverricht has described a form of myoclonus, occurring as a family disease and associated with epilepsy. In addition to the general clonic movements, the tongue, the muscles of deglutition, and even the diaphragm may be involved. The epileptic seizures may be few and far between in earlier years, are often nocturnal; later on epileptic attacks occur more frequently. Lundborg, Clark and Prout have given especial attention to this association disease. (See Clark and Prout's article, American Journal of Insanity, No. 2. 1902.)

CHAPTER VII.

TETANUS.

In view of recent bacteriological researches tetanus may be described as an infectious disease of the central nervous system, characterized by continuous tonic spasms with occasional attacks of clonic movements. The muscles first affected are, as a rule, those of the neck and jaw. Opisthotonus and lock-jaw (trismus) are the first and most startling symptoms of the disease. According to its origin we distinguish between traumatic tetanus resulting from a wound, and idiopathic or rheumatic tetanus if the cause is unknown, or if, in our ignorance, we attribute it to exposure to wet and cold. We must take special note of the tetanus occurring in new-born children (tetanus neonatorum), and may make a passing reference to puerperal tetanus. There is every reason to believe that the tetanus bacillus, discovered by Nicolaier and developed in pure cultures by Kitasato, or its toxic product (studied by Brieger chiefly) is the direct cause of tetanus.

Etiology.—Tetanus is far more frequent in males than in females. This is particularly true of those cases in which the traumatic factor is evident, and is easily explained by the greater liability of the male sex to traumatism. The very fact that of 46 so-called idiopathic cases recorded by Gowers only 9 were in females would lead one to doubt the spontaneous origin of these cases. While tetanus may occur at any time of life, it is most frequent between the ages of ten and forty years. It occurs within the first few days of life between the fourth and eleventh days, but between this time and the age of five years there is, practically, immunity. Between five and ten years the tendency to the disease is on the increase, and continues to increase until after the age of forty years. These statistics strengthen

the impression that traumatic lesion alone, plus infection, is the only feasible explanation of the occurrence of the disease.

Colored races, and particularly those living in tropical countries, are affected more frequently than white people. This has been amply proved in regard to the population of India and the West Indies. The filth and uncleanly habits, so common among these people, should be borne in mind. Puerperal tetanus, which is unquestionably infective in character, occurs very frequently still in these same countries, but it is becoming less and less frequent in those countries in which modern surgical principles prevail in obstetrical practice.

The previous health of the person seems to have little to do with the development of the disease. The most robust as well as the feeblest are equally liable to the infection. Irregular wounds, burns, scratches, and frost-bites are more often the cause of tetanus than the large aseptic incisions made by the surgeon's knife. Tetanus has been known to follow the extraction of a tooth, and, in the popular belief, lock-jaw is much feared after slight injuries to the toes in paring nails, etc. With greater cleanliness among the masses the disease will no doubt soon diminish. Thamhayn states, from a large collection of cases, that tetanus is developed most frequently from wounds in the hand, leg, foot, head and neck, arm and trunk, in the order of frequency just mentioned. Injuries to the head have often been supposed to be peculiarly liable to produce tetanus; but as such injuries are generally the result of a fall and contact with the earth, as in a fall from a horse, it will be seen that the danger of infection is greater than in many other instances. The same is true of tetanus following a splinter which has been run under the nail, and the old-fashioned method of stopping bleeding by putting cobwebs around the bleeding part may be very largely responsible for the tetanus. The micro-organisms of tetanus evidently do not require a large abraded surface, and it is questionable whether, even in those cases in which tetanus comes on after flogging, there was not sufficient abrasion of the epithelium to permit of the invasion of these minute organisms.

As a rule, tetanus is developed within a period of five to fourteen days after the initial injury; but, in some cases, a few hours are sufficient for the development of the disease, and in cases in which the crural nerve has been enclosed by a ligature, full-fledged tetanus of the severest type was observed at once after the occurrence. The development of tetanus is not rare within three or four weeks after the injury, but beyond such a period the occurrence of tetanus could not well be explained. That infection alone will not explain all the circumstances in connection with the development of tetanus will be insisted on in the section on pathology.

SYMPTOMS.—The first symptoms of tetanus, if we except vague pains at the seat of injury, and a dull headache, is a gradual stiffening of the muscles of the neck and of the jaw until the head is markedly retracted (opisthotonus) and the jaws are firmly clenched (trismus). While the cervical muscles are apt to be involved at the beginning the dorsal and lumbar apparently escape; but retraction of the head is soon followed by arching of the back. As the disease continues the legs become rigid, but the arms escape or are but little involved. The muscles of the face are often affected at an early stage of the disease. The eyebrows are raised, the ocular fissure becomes narrow, the mouth is distorted, the lips press against the teeth and the entire face may assume what is known as the "risus sardonicus." The rigidity of the muscles is painful, and the occasional clonic exacerbations which occur tend to make the condition still more distressing. The difficulties of deglutition and of respiration, from spasm of the thoracic muscles and of the glottis, increase the agony of the patient and are a continuous menace to life. Epigastric pains (pains possibly due to spasm of the diaphragm) are frequently complained of. During sleep the spasm usually ceases, but no sooner does the patient wake up than all the distressing symptoms return in full force.

The pulse is rapid and feeble. Whether vaso-motor spasm is the cause of this peculiarity of the pulse has not been sufficiently determined. The temperature varies considerably; in some instances it remains normal during

the entire course of the disease, in others there is a continuous fever of from two to three degrees. H. C. Wood refers to the serrated tracings of the temperature chart, the rise coinciding with the paroxysm, and the fall of the line with the interval. Occasionally the temperature rises to 108° or even 110° F., and according to Wunderlich this high temperature continues for some hours after death, reaching even to 114° F. It was formerly supposed that the rise of temperature was due to excessive muscular action; but cases in which muscular action is greatest are not necessarily those with highest or even high temperature. Gowers is inclined to consider the fever to be of nervous origin, which is probably correct if modified to mean that the toxines circulating in the system exercise a special irritating influence upon the centres in the pons and spinal cord, or upon the heat areas in the cortex. Thirst is a frequent and most distressing symptom of the disease, and as it cannot be easily quenched, on account of the difficulty of deglutition, it adds much to the discomfort of the patient. The thirst is also increased by the very profuse perspiration which sets in and is a prominent factor in many of the cases.

The urine is scanty and high-colored, of high specific gravity, all of which can be explained by the excessive action of the skin. Micturition is often irregular, possibly in consequence of spasm of the sphincter. The bowels are, as a rule, constipated.

All the symptoms are apt to increase in intensity as the disease progresses. The majority of cases run to a fatal termination in less than a fortnight, often in four to five days. Death occurs, as a rule, either from failure of the heart or from asphyxia in consequence of spasm of the respiratory muscles. In other cases exhaustion from the continued spasms and from the inability to take food may be the ultimate cause of death. Cases lasting above a fortnight are supposed to tend toward recovery. The spasms become slighter, the clonic spasm less frequent, and all the symptoms gradually recede until nothing but a slight rigidity of the muscles remains. The parts last involved are first released, and the spasms last longest in the muscles of the

neck and jaw, the very parts which were first affected. A single recovery seems to provide immunity against the disease for all time. While the evidence upon this point is not absolutely conclusive, the fact of immunity after the first attack would be in keeping with what we know about immunity from other infectious diseases.

VARIETIES.—The majority of the cases are so much alike that it would be quite useless to speak of variations from the clinical type. An exception must be made, however, in favor of what Rose has termed "cephalic tetanus." This form develops, as a rule, after injury to the head, and particularly to the region of the face supplied by the fifth nerve. The special characteristics of this form are the association of paralysis of the face on the same side as the injury with tetanic spasms of the other side. In addition to this we find spasms of the respiratory muscles and great difficulty in deglutition. The behavior of the patient is very much like that of one suffering from hydrophobia, hence this form has also been spoken of as "tetanus hydrophobicus." There is some doubt whether injury to the peripheral nerve is the cause of the facial palsy. Bernhardt and Remak proved conclusively that there was no reaction of degeneration in the nerve or muscles, and the nerve has been found healthy on post-mortem examination. We must suppose, therefore, that the facial palsy in such cases is of reflex origin. Some support is given to this view by the fact that if recovery sets in, the tetanic spasm and the facial palsy disappear at about the same time.

The only other variety worthy of special mention is the so-called abortive form of tetanus described by Kussmaul. In these cases spasm of the neck muscles and trismus are the only symptoms, and these gradually disappear.

Tetanus neonatorum can hardly be considered a variety of tetanus, for it resembles in every way the tetanus of the adult, but in this book it will naturally deserve some special consideration.

The tetanus of the new-born child sets in, as a rule, between the fifth and tenth days after birth, though it may be delayed as long as twenty days. The first symptom noticed in the child is difficulty in taking the nipple or in drinking

from the bottle; with every attempt there is distinct rigidity of the masseters and of the muscles about the mouth, which interferes with the act of sucking and swallowing. Other facial muscles are apt to be in a state of contracture leading to a distinct deformity of the face. In some instances food can be poured into the mouth by a spoon without exciting a spasm, but in many others the mere touch of the spoon to the lips, or contact of the food with the lips and mouth, is sufficient to produce a spasm.

In the earlier stages of the disease the child is quiet unless food is given; but before long the spasms come on spontaneously without excitation by food. Spasm of the respiratory muscles soon forms a part of the clinical symptoms, and periodic cyanosis is often one of the early symptoms of the tetanic condition. Rigidity of the muscles of the neck and back, typical opisthotonus, and arched back soon follow, and to make the symptoms thoroughly complete the upper and lower extremities become the seat of muscular spasms. There may be slight remissions for a time, but the tetanic stage becomes more and more permanent, or, if absent when the child is quiet, will surely come on with the merest touch in lifting the child, in trying to give it an enema, in washing it, etc. The temperature, as a rule, remains normal; in a few instances it reaches 102° or 103° F.

The disease is steadily progressive. One is often deceived by slight remissions and is flattered by the temporary good result of sedative treatment, but is only too often disappointed by the reappearance of all the symptoms, with even greater intensity. Death may follow within a period varying from one and a half to eight or nine days, and results either from mere exhaustion, due to the inability to take food, or from asphyxia, due to spasm of the inspiratory muscles.

A few cases of tetanus neonatorum get well, but surely only a very few. Cases with fever seem to be invariably fatal. In those who get well the tetanic spasm disappears very slowly, and Henoch states that in some instances a rigidity of the muscles existed three weeks after the onset of the disease.

The cause of this early tetanus is in every way the same as that of tetanus in the adult, except that the entrance of the bacilli into the body occurs by way of the umbilical cord, the care of which has been neglected. In former days, and particularly in those countries in which obstetrical cases were managed exclusively by midwives, whose ideas of cleanliness were very defective, tetanus neonatorum was very much more frequent than it is now. Ritual circumcision, if performed without due regard to modern surgical principles, may be considered an occasional cause of tetanus. Bathing a child at low temperatures is referred to by many of the older writers as a common cause of tetanus. The use of a bath thermometer is said to have put an end to an epidemic of tetanus occurring in the practice of a midwife who was not able to distinguish by the hand alone the proper temperature of water. In view of the modern bacillary theory of tetanus such an origin of tetanus would be rather difficult to explain; but we must concede the possibility of other than mere toxic agents affecting and irritating the peripheral nerves of a newly born child.

Pathological Anatomy and Morbid Pathology.—Careful search has been made by many writers for the actual lesion of tetanus, but up to the present time with few positive results. It has been noted that rigor mortis sets in with unusual promptness, some claiming that the tetanic spasm passes without relaxation into the post-mortem contracture. All the organs of the body have been examined; no special changes have been found in them. Some writers have reported œdema, hypostatic pneumonia, pleural extravasation, which can be accounted for by the interference with the circulation and respiration. The muscles of the body are sometimes ruptured; some contain small hemorrhages, but under the microscope the fibres present no anomalies, though Bowman refers to occasional granular degeneration. It has been natural to look for changes in the nerve filaments in the peripheral wound which has been the starting-point of the tetanus. In some instances nothing abnormal has been found; in others there has been neuritis, with considerable swelling of the nerve, which extended from the periphery to the spinal cord.

In the case of tetanus neonatorum an inflammation of the umbilical vessels has been traced for some distance within the abdomen, and the peritoneal covering of the vessels has also been found inflamed.

As for the central nervous system, the condition most frequently discovered is one of passive distention of the vessels, associated with minute hemorrhages; but both these conditions are surely secondary to the tetanic spasms and not in any sense the cause of them. In the spinal cord some investigators have reported granular disintegration of the gray substance, the formation of cavities containing granular material, and changes also in the large ganglion cells, such as were found by Nerlich in the motor root of the fifth nerve. All of these changes are not sufficiently distinct to represent the actual pathological conditions of tetanus; they are secondary effects, and it is probable that in tetanus, as in other of the diseases hitherto considered, definite though the symptoms may be, the changes are of a transitory character, every trace of which disappears after death. It would be more important, therefore, to refer to the morbid pathology.

FIG. 49.—Tetanus Bacilli and Spores. (× about 1,000.) (Kitasato.)

The *pathology* of tetanus can be readily understood in the light of modern researches if we regard the spasms of tetanus as the result of increased irritability of the convulsive centres in the brain. This increased irritability is directly due to the influence of germs or germ products introduced into the system from some external injury. The tetanus bacillus (Nicolaier) is a rod-shaped microbe whose spores are attached to one end of the bacillus, which with its spores resembles in appearance an ordinary pin. This microbe is found in the soil as well as in the dust of dwellings. These bacilli are capable of resisting great heat,

retaining their activity even after exposure to 175° F. for an hour (Kitasato, *Zeitschrift für Hyg.*, vol. vii.). Their spores are capable of resisting the same heat for six hours, but the spores are not formed if the temperature exceeds 108° F. The bacteriologic phase of this subject has been studied most carefully in recent years by Ehrlich, Wasserman and Milchner, to whose writings the reader is referred.

As products of these bacilli we have several poisons (tetanin, tetano-toxin, and also *tox-albumins*, Brieger), which have been derived from pure cultures and which are capable of exciting the disease. It is probable that the tetanus bacillus itself does not cause the disease, but that the poisonous substances formed in the blood by the presence of this bacillus are the direct cause of tetanus. It is an interesting fact that granulating surfaces do not offer a suitable soil for the production of these toxic substances. The presence of oxygen seems to prevent the penetration of the bacilli into the tissues underneath; hence we can readily understand why scratches and hidden injuries should be far more dangerous than large abraded surfaces.

Tetanus was supposed to result from direct irritation of the peripheral nerves; if so, an ascending neuritis would have to account for the development of the symptoms of tetanus. It is at present altogether impossible to deny that in some way or other the bacillus must have been introduced into the system even in these cases.*

Lesions of the occiput were supposed to lead more frequently than other lesions to tetanus, but in this instance, too, it is more than probable that the infectious element cannot be disregarded.

In no disease have bacteriological researches led to more satisfactory results in treatment. Tizzoni cultivated the virus, inoculations of which in increasing strength produced tetanus in dogs. The blood-serum of such dogs destroyed the activity of the virus. He inferred the pres-

* Since the above was written Goldscheider has given a different explanation based upon recent anatomical conceptions. The poison of tetanus acts upon the peripheral nerve-fibre. This fibre being the peripheral portion of the neuron, an increased excitability of the ganglion cell is produced, and this hyper-excitability brings about the tetanic contractions of the muscles with which it is connected.

ence of an albuminoid body, which he called "anti-toxin," with which he succeeded in arresting the disease in rats.

The now famous researches of Behring and Kitasato have revealed the fact that the blood-serum of tetanic animals produces immunity in others, and that the serum of the animals thus rendered immune has still more active anti-toxic qualities. It was during the year 1894 that Behring showed that the blood-serum of immunized animals not only produces immunity in others, but also has a distinctly curative effect. Tarnuffi (*Centralblatt für Bacteriologie*, 1892) reported a successful use of anti-toxin injections in the case of a peasant, aged seventy-four, who had developed tetanus as the result of a lacerated wound of the little finger of the right hand. One hundred and fifty grammes of the anti-toxine was given in this case, when on the eleventh day after the beginning of the injections a complete recovery ensued. The blood-serum from this patient did not produce tetanus when injected into a rat. No doubt other successful cases of this kind will soon be reported, and there is much to hope from this serum therapy in the case of tetanus.

Differential Diagnosis.—The diagnosis of tetanus is easy in the majority of cases. It is possible to mistake tetanus for strychnia poisoning, but in the latter the symptoms never begin with distinct trismus. The symptoms also develop with much greater rapidity, and the severe pain referred to the stomach, as well as the absence of external injury, would lead one to suspect the effect of strychnia.

If there is great difficulty in deglutition the symptoms of tetanus may resemble somewhat those of hydrophobia, but the entire absence of trismus, and the fact that all the symptoms are excited only if an attempt is made to swallow, and the previous history of the case, will enable one to distinguish hydrophobia from true tetanus.

As for mistaking tetanus for tetany, such confusion is scarcely possible if the mode of onset from the periphery inward, in cases of tetany, is kept in mind, and if we also remember that no such interval exists in tetanus as in the lesser disease. The favorable course of the disease will

render still further aid, after the lapse of a few days, in differentiating between tetany and tetanus.

PROGNOSIS.—The prognosis is invariably grave. A mortality of eighty-five per cent. does not overstate the facts. Other things being equal, the prognosis is the more grave the more thoroughly a lacerated wound has been infected. Recovery in a case of tetanus after compound fracture of the limbs is very rare. The longer the interval between the injury and the first onset of the symptoms the better the prognosis, and cases which begin after ten days usually take a more favorable course than those which come on very early after the injury. If the symptoms have lasted more than ten days there is some reason to expect a favorable result. On the whole it is a disease very much to be feared, and a guarded prognosis is quite in order until a very decided improvement sets in ; but possibly the recent advances in serum therapy may in the course of time enable us to be more hopeful.

TREATMENT.—In the treatment of tetanus, prophylactic measures are by far the most important. There is no good excuse at the present day for the infection of any surgical wound, or if the wound has been received under conditions which make infection possible, thorough antiseptic measures should be at once employed, and even after the first symptoms of tetanus have been observed the condition of the peripheral wound should be carefully examined into and everything should be done to prevent further contamination and infection.

If recent investigations are carried to a successful issue, the most satisfactory and the most rational treatment of tetanus will unquestionably be that by injections of antitoxin or any of the substances which may hereafter be proved to possess the property of counteracting the poison of tetanus in the human system. Behring has perfected the anti-toxin treatment of tetanus, insisting, also, that the injection be made as near as possible to the seat of infection.

In the majority of cases, and particularly in those occurring at a distance from large medical centres, from which for years to come these anti-toxins will have to be obtained, the older methods of treatment will still have to be resorted to.

The general management of the patient is the first important consideration. Absolute quiet should be secured by darkening the room, keeping out all visitors and noises. Nutritious but liquid diet should be given, and given slowly, so that the difficulties in deglutition may not be increased. If the trismus is very marked a wedge may have to be inserted between the teeth to secure enough space to introduce a tube through which liquid can be taken into the mouth. If this cannot be done, the patient must be fed through the nose or else by the rectum.

In former times it was considered wise to remove a peripheral scar, or even to amputate the part that included the seat of injury; but according to our present conception the presence of tetanus bacilli in the system would prove that the disease was no longer a local one, and that excision of the lesion would not prove sufficient to bring about recovery.

Among the drugs employed in the treatment of this disease chloroform by inhalation is unquestionably the most satisfactory. Woods, of Philadelphia, has recommended the use of nitrite of amyl, and Gowers reports that its use at Guy's Hospital has proved that the spasms became more intense at first but slighter afterward. Both chloroform and nitrite of amyl are palliative measures, but not curative in any true sense. Chloral hydrate has an advantage over the preceding drugs, inasmuch as it helps to produce sleep, and can be given continuously for a long period of time. To be effectual large doses of five to fifteen grains should be given at a time, and repeated several times during the day according to the needs of the case.

There can be no reasonable objection to the use of opium and morphia in the form of suppositories, or by hypodermatic injection. Morphia may be exhibited in doses of one-twelfth to one-sixth grain, according to the age of the child, and can be given in connection with other narcotics. The use of conium and of gelsemium has been warmly recommended by various observers, but the results are not satisfactory enough to advise the general adoption of such measures. Curara in doses of one-fiftieth to one-half grain every hour has been urged by some; but it is

surely a dangerous drug, and its application would be rather in the nature of an experiment than of safe therapeusis. Continued warm baths, electricity (galvanism to the spine and the muscles), all these measures have been advocated by some, but without much result. If the bromides, chloral, opium or morphia, and chloroform, judiciously administered, do not bring about a favorable result it is safer not to exhibit other drugs.

In the treatment of tetanus neonatorum the same drugs may be employed as are recommended in tetanus of the adult, but the dosage should be modified in keeping with the age of the patient.

CHAPTER VIII.

TETANY.

TETANY or tetanilla is a disease which occurs with far greater frequency in Europe than in this country. Osler and Weir Mitchell speak of its exceeding rarity in the United States. It occurs with about equal frequency in adults and children. As the name indicates, "the little tetanus" is characterized by attacks of tonic spasms of various groups of muscles, particularly of those of the upper extremities. It is only within recent years that the disease has been properly studied, though the first account of it goes back as far as the year 1830, when Steinheim described this group of symptoms as a special form of articular rheumatism. In the following year Dance published an article in which he expressed the view that the intermittent character of the spasms proved the disease to be of a malarial order. The name of the disease we owe to Corvisart, who in 1852 reviewed the entire subject. His observations were preceded, however, by those of Trousseau, who had observed the disease in nursing women, and assumed a connection between tetany and the function of lactation. He therefore termed it "contracture rheumatismal des nourrices," but later on he observed the trouble in children and in adults with intestinal obstruction, and so abandoned the lactation theory. It was Trousseau who first discovered the very important fact that these attacks could be excited by compression of the arteries and nerve-trunks of the affected extremity.

Our knowledge of this disease has been enhanced by the observations of Chvostek, Koppe, Baginsky, Von Jaksch, Bernhardt, and Escherich. The most comprehensive articles on the subject have been written by Weiss, of Vienna, in 1881, by Frankl Hochwart and by Escherich.

SYMPTOMATOLOGY.—The symptomatology of tetany includes the symptoms to be noticed during the attack and those during the period of latency. The attack is preceded

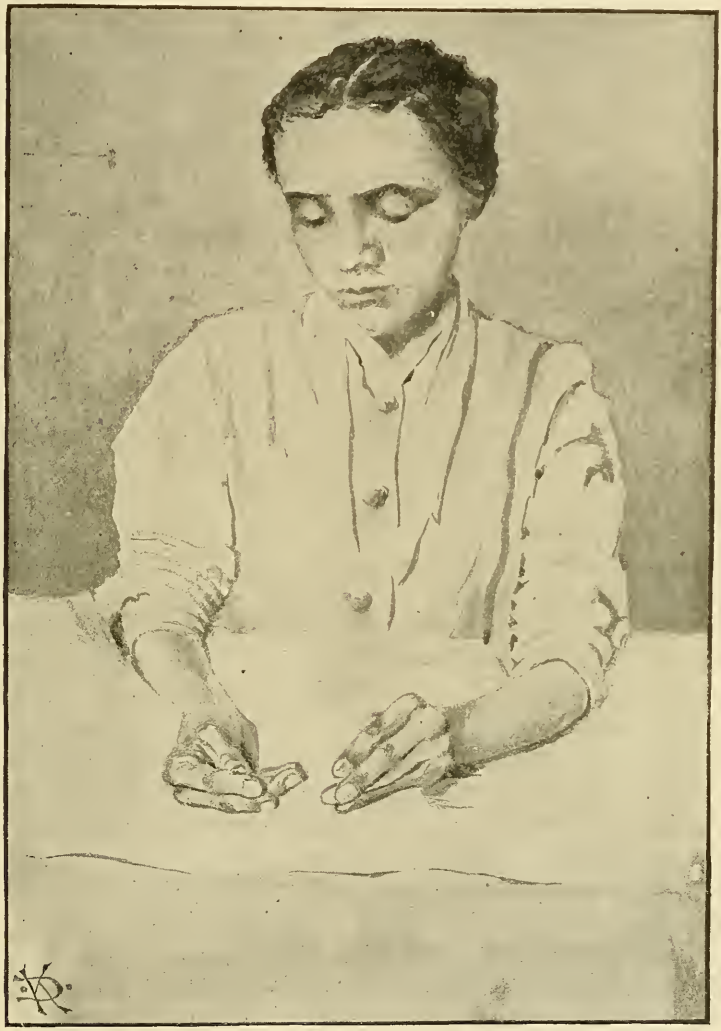


FIG. 50.—Position of Hands in the Spasm of Tetany. (After Oppenheim.)

by vague tingling pains, by formication in the hands, forearms, and legs; a feeling of stiffness soon follows, and subsequently the spasm of the muscles sets in. This tonic spasm

occurs more frequently in the upper extremities, and gives rise to such a marked rigidity of the muscles that passive movements are for the time being impossible. The position of the hand during the spasm will necessarily vary according to the groups of muscles affected. It is a common occurrence for the hand to assume the shape of the accoucheur's hand. Occasionally also the thumb is very firmly pressed upon by the flexed fingers and the nails are buried in the skin of the palm. In some rare cases there is complete extension of all the fingers. The forearms are generally flexed, and the upper arms closely pressed against the chest. If the lower extremities are involved the thighs may be adducted, the legs extended or flexed, while the toes are apt to assume the position of talipes equinus. The spasms may also affect the muscles of the abdomen, of the chest, and of the back. These tonic contractions of the abdominal and thoracic muscles may interfere with the movements of the diaphragm, and with the respiration, causing dyspnœa and cyanosis. If the muscles of the neck be also involved, the return of the venous blood from the brain may be retarded, as reported by Weiss in one case, in which loss of consciousness was the result of such neck spasm. Opisthotonus is not infrequent, but trismus is rare and never occurs in the beginning of an attack as in tetanus. In severe cases spasms of the ocular muscles have been observed, of the œsophagus, of the larynx, and of the muscular apparatus of the bladder (a desire to urinate but micturition impossible). During the attack the patient complains of severe pain in the affected muscles; there is also a diminution of tactile sensibility in the extremities, the muscular sense is often deficient, and while standing on the floor patients have a feeling as though they were walking on velvet.

Elevation of temperature as high as 104° F. has been observed; but this is exceptional, as Weiss records an average of only one such case in twelve.

Headache, vertigo, tinnitus aurium, and excessive perspiration are other symptoms which have been noted during an attack of tetany. The attacks may last only a few minutes, or may cover a period of hours, or even days. Severe attacks of tetany may bear a striking resemblance to

genuine tetanus; but it should be noted that there is no initial spasm of the masseters in tetany, and that in this form the spasm spreads from the periphery inward, and not centrifugally, as is the case in tetanus. Moreover, the reflex excitability, is not nearly as great in tetany as in tetanus. In tetany the patients may be entirely free from attacks for hours, and even for days, whereas in tetanus the symptoms, as long as the attack lasts, are continuous.

Etiology.—The disease occurs chiefly in very young persons. The cases observed by Koppe were all from one to two years of age. Ganghofner reported 40 cases; of these 5 were between two and three years of age and the remaining 35 between the ages of one month and two years. Gowers tabulated 142 cases; of these 64 occurred from one to four years, and 36 from ten to nineteen years of age. On going over the literature of the subject a few years ago, taking all cases of tetany into consideration, it was evident that the majority of cases of tetany were observed between the ages of sixteen and thirty-five years. At one time the disease was classed among the professional neuroses, but this fallacy was corrected by Kussmaul. Any exhausting disease may be regarded as a possible etiological factor, but exposure to cold and wet is referred to most frequently as a predisposing cause. Intestinal irritation is another cause, and may be associated with eclampsia and laryngospasmus, two conditions which we know are also frequently excited by intestinal irritation. Riegel instances a case in which attacks of tetany were inhibited by the removal of the ova of tænia medio-canellata and tricocephalus dispar. Weiss reports the occurrence of tetany as a complication of typhoid fever. It has also been observed together with small-pox, Bright's disease, malaria, chorea, and even after severe mental shock. The only just inference from all this is that tetany is liable to occur after any exhausting disease in those who are predisposed to this form of spasm. Weiss deserves special credit for bringing into prominence the relation between tetany and extirpation of the thyroid gland. The disease sometimes appears in epidemic form, if we are to credit the account of such occurrences in the schools and prisons of France. A similar epidemic occurrence is re-

ported by J. Lewis Smith, Escherich, and others. That it is much more frequent in some countries than in others has already been alluded to.

The male sex is affected a little more frequently than the female; Rilliet and Barthez recording 20 out of 28 cases in boys. In Gowers's statistics of 142 cases, 76 were males and 66 females. Although the disease sometimes occurs in families there seems to me to be insufficient evidence of actual hereditary predisposition to this disease.

Symptoms of the Latent Period.—In the interval between the attacks the patient may appear to be entirely well, but if examined carefully he will exhibit a weakness, with slight rigidity of the affected muscles. The calf muscles are commonly the seat of mild contractions. Chvostek observed slight contractions of the orbicularis palpebrarum.

The intervals between the attacks of tetany may vary in duration from several hours to a few days, and even a few months; but, of course, we can speak of a latent interval only in case the disease can be proved still to exist. This can be done by eliciting Trousseau's symptoms and by proving an increased electrical and mechanical excitability of the parts affected.

Trousseau's Symptom.—The famous French physician discovered that in persons afflicted with tetany a characteristic attack can be elicited by pressure upon the large nerve-trunks and the arteries of the extremities usually affected during an attack. The attack ceases as soon as pressure is removed. Kussmaul and Quincke maintain that in some cases pressure on arteries only is necessary, while in other cases the slightest pressure on a nerve-trunk is sufficient to produce contractions of all the muscles supplied by this nerve. Trousseau's symptom is absolutely pathognomonic of this disease.

Increased electrical excitability has been found to be characteristic of tetany by Erb, Chvostek, Weiss, and others. According to these authors the faradic and galvanic responses of the motor nerves are enormously increased during the interval between the attacks of tetany. They were not only able to obtain kathodal closure contractions (K.

C. C.) with very small currents, but were able with moderate currents to obtain a kathodal closure tetanus, and even an anodal opening tetanus, which has not been observed in any other condition. Chvostek and Weiss claim that this phenomenon is exhibited in the facial and in other peripheral nerves. Erb found the electrical excitability greatest at the time when the attacks were most frequent.

Increased mechanical excitability is a still more striking symptom of the condition, a simple touch with a percussion-hammer upon a nerve-trunk being sufficient to produce contractions of the muscles supplied by the nerve. I remember distinctly the cases in the Vienna General Hospital in which pressure with a lead-pencil upon the focal point of the pes anserinus was followed by contractions exactly like those which a strong faradic current applied to this part would have produced. This is by far the most convenient test to make in cases in which the existence of tetany is suspected, and it is far better to endeavor to establish this fact of increased mechanical excitability than to excite an attack by pressure upon a large nerve-trunk or a large artery.

Differential Diagnosis.—There can be but little difficulty in differentiating between tetany and other convulsive disorders. From tetanus it can be distinguished sufficiently by the intermittent and centripetal character of the attacks, by the absence of trismus at the beginning of the attack, and, above all, by the presence, during the interval, of Trousseau's symptom, and the increased mechanical and electrical excitability. I have known a case of frequently repeated epileptiform convulsions to be mistaken for a case of tetany, but the loss of consciousness, the universal tonic and clonic movements, the turning of the eyes, the stupor following an attack, and the extreme shortness of the attack, are the symptoms that will help to differentiate the epileptiform attack from one of tetany.

Morbid Anatomy and Pathology.—Post-mortem examinations of cases of tetany have been made by Langhans and Weiss, but up to the present day there have been few positive pathological findings. Langhans claims to have found a periarteritis and periphlebitis of the white commis-

sure and of the anterior horns of the cervical portion of the spinal cord. Weiss failed to find any such changes in his cases. He formulated an ingenious theory of the disease according to which the attacks of tetany are due to an irritable condition of the gray matter of the medulla and spinal cord. This irritable condition is the result of sympathetic disturbances causing irregularities in the vascular innervation of the blood-vessels of the spinal cord. Gowers is inclined to look for the chief changes in the motor cells of the spinal cord, but all this is in the nature of speculative pathology. Schlesinger has very recently reviewed the various theories concerning the nature of tetany. He concludes that none of them is entirely satisfactory, while, according to his own ideas, tetany is a disease of the entire nervous system, some of the symptoms being due to an affection of the peripheral nerves; but the tetanic spasms, as well as Trousseau's phenomena, are held to be the result of increased excitability of the gray matter of the central nervous system, chiefly of the brain, medulla oblongata, and spinal cord. Peripheral irritants of various kinds may give rise to vasomotor disturbances in the spinal cord, and these may be the cause of functional changes. This last theory is but a little more satisfactory than the preceding ones, for nothing is known as yet of that special peculiarity and special irritability of the central nervous system which give rise to this disease. Irritability of the nervous system is so common that something more is needed to explain these very unusual manifestations of the disease. That a zymotic factor enters into the etiology of tetany can scarcely be doubted, but the exact nature of such infection is a matter for future study.

The occurrence of tetany after extirpation of the thyroid gland points the way to a future investigation regarding the origin of the disease. If it is the function of the thyroid gland to eliminate mucin from the body, it is natural to infer that the continuance of this toxic substance in the system is responsible for the tetanic spasm that sets in when the gland is removed by surgical interference.

A special caution is in order not to confound ordinary carpopedal spasm, or any of the short clonic and tonic

spasms that so frequently occur in children, with true tetany.* The term should be restricted to those cases that present distinct attacks and a free interval, characterized by Trousseau's symptom and increased excitability.

PROGNOSIS.—The prognosis is favorable excepting in those few cases in which serious lung trouble may result from continued spasm of the respiratory muscles; but the prognosis should be very guarded with respect to the duration of the disease. It may vary from a few weeks to many months.

TREATMENT.—The removal of every possible peripheral irritation is the first *sine qua non* of treatment. If intestinal irritation is suspected to be the cause, free purging of the bowels and removal of intestinal parasites are called for. A change of abode may become necessary, and absolute rest will have to be secured at any cost.

To shorten the attack the physician will probably have to resort to the hypodermic use of morphine, and possibly to hyoscine. Weiss reports a single case in which the attack was inhibited by the application of ice to the back of the neck. Gowers, who seems to think the spasms of tetany very closely related to ordinary infantile convulsions, advises treatment similar to that employed in convulsions. Proceding on this basis, inhalations of nitrite of amyl and of chloroform may be tried. As soon as the attack is over it would be well to administer chloral hydrate in daily doses of about one-half drachm, or the bromides in doses of from ten to twenty or more grains per day, according to the age of the child. Small doses of sulfonal, or of trional, are worthy of trial. During the interval careful electrical treatment (stabile ascending currents through the peripheral nerve-trunks) as well as prolonged lukewarm baths should be given. There is little doubt but that the majority of cases will get well without any therapeutic measures. The chief duty that devolves upon the physician is clearly that of invigorating the central nervous system by the best known methods and remedies, in order to enable it to resist the ordinary irritants which produce the disease in those who are predisposed to it. If the child exhibits any symptoms

* Some of the cases reported by Vaughan are not distinct cases of tetany.

of rickets, cod-liver oil and iron will be the best remedies, and treatment, to be successful, will have to be directed entirely to the improvement of the child's defective bone development. The bromides may be administered if the condition is not intense enough to call for narcotics and opiates, and if, for some reason or other, these latter remedies should not be employed. Gowers states that a dose of digitalis, given at bedtime, has been found to be the most useful remedy for nocturnal tetany. The action of the digitalis could be explained only on the theory that the blood-supply of the central nervous system is deficient, and that there is need of increased activity.

TETANOID CHOREA AND OTHER TETANOID DISORDERS.

Under this title Gowers refers to a case which has been under his care that exhibited symptoms intermediate between those of chorea and that of tetany. The disease ran a fatal course, but no demonstrable lesions were found after death. The patient was a boy, ten years of age, with a history of three other relatives having suffered from diseases resembling chorea. In the patient the symptoms began seven months before death. They consisted of tonic spasm, which was continuous, and varied by paroxysmal attacks of similar but more intense spasm. The face was involved on both sides so as to cause a constant peculiar smile. The tongue was pressed up against the palate, impeding swallowing and preventing speech. The arms were extended, pronated, and rotated inward so as to bring the back of the forearms outward, while the fingers were flexed at the joints, but at times were extended and moved slowly in a way characteristic of athetosis. The lower extremities were extended at all the joints, the feet being extended in talipes equino-varus, and the toes were flexed. The muscles of the trunk were also involved in the spasm. At first the left side was the more severely affected, but afterward the spasm became equal on the two sides.

The electrical irritability of the muscles was normal, and there was no mechanical excitability of the nerves. There was considerable pyrexia during the more severe stage of the disease. The boy became thoroughly emaciated, and died from exhaustion. There is very little of true tetany, and still less of chorea in this case as described by Gowers. While I have seen no case exactly like this one of Gowers, it is not rare to find irregular spasmodic movements of all sorts that may remind one at times of chorea, at times of athetosis, and then again of a cataleptic condition during febrile disorders in children.

Escherich, of Vienna, who has been the most recent and a most careful student of tetany, calls attention to the great frequency of tetany and of tetanoid disorders in young children. Of eighty cases of tetany which he

observed in the Children's Hospital, the majority occurred between the ages of three to thirty months and he is inclined to infer that the artificial nourishment given to these children is largely responsible for the development of the tetany. Fully one per cent. of all the children under three years of age which have been treated in Graz were suffering from tetany. It is safe to say that such frequency is peculiar to certain cities. It is surely very different from the experience of American pediatrists and neurologists. I wish specially, however, in this connection to allude to Escherich's conclusions that even more frequent than tetany are various tetanoid disorders in the earliest years of life. This author goes to the extent of putting laryngospasm, muscular spasms and eclampsia into a series characterized by those signs which would lead one to infer that all of them were tetanoid in character. The present writer has been on the look-out for this special combination of conditions and he is confident that in America this combination is not a very common one. In only a single instance has he seen a child that presented the symptoms of laryngospasm that was in a condition of muscular contracture resembling that of tetany and that at times had distinct convulsive seizures. In such cases the tetanoid character of the condition is proved by the occurrence of Trousseau's and Chvostek's symptoms either at the time of the spasm or during the period of latency. Escherich's observations should encourage us to look for the existence of a latent tetany. I am confident, however, that these conditions are far less common in America than they are in Austrian and French cities.

CHAPTER IX.

HEADACHES.

It requires no little skill on the part of the physician to discover the true cause of headaches. They do not constitute a form of disease, but are symptomatic of many organic and functional conditions. For this reason we must give them the very closest study. The diagnosis of the kind of headache in the adult is difficult enough, but it is doubly so in the case of children. As the headaches that come on in early life are frequently continued in later years and give rise to much suffering, it is particularly important that an effort should be made to nip them in the bud. The most distressing form of headache and the one most stubborn in its resistance to treatment, migraine, will be considered in a separate section. The remaining forms may be classified as follows:

Headaches due to:

1. Anæmia and malnutrition.
2. Neurasthenia.
3. Transitory hyperæmia, as it occurs in heart disease or at the period of menstruation.
4. Gastric disturbances.
5. Genital irritation.
6. Ear disease.
7. Organic disease of the brain or its coverings (including specific disease).
8. The prodromal stage of acute infectious diseases.
9. Malaria.
10. Uræmic poisoning.
11. Other toxic conditions.

It was the fashion formerly to make a differential diagnosis of the forms of headaches according either to the

character of the headache or to the seat of the distribution of the headache. Thus frontal headaches were considered to be due to gastric disturbances. Occipital headaches were considered to be almost pathognomonic of uræmic poisoning; but he who has an opportunity of seeing a large number of cases of headaches in the adult, as well as in children, will soon convince himself that these signs are often misleading. I have seen frontal headaches with disease of the kidneys, and occipital headaches from gastric disturbances and in cases of specific disease, of tumor of the brain, etc. The diagnosis of the kind of headache can, as a rule, be made after carefully examining into the general health of the patient,

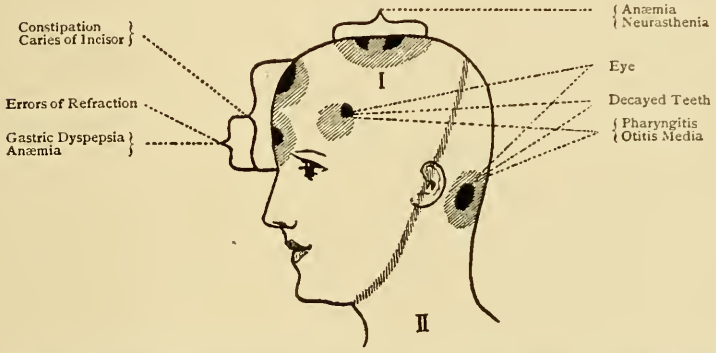


FIG. 51.—Location of Head-pains. After Dana (slightly altered). Area I. Trigeminus and Facial Nerve Strands. Area II. Upper Four Cervical Nerve Strands.

and endeavoring to find what the fundamental disturbance may be. It will be better for us, therefore, to take the etiological conditions mentioned above, and to endeavor to connect with each condition a few signs that are generally associated with a particular form of headache.

1. Headaches due to anæmia. These headaches occur frequently in children between the ages of eight and fifteen years; they are sometimes frontal, generally vertical, and are described, as a rule, as a dull, boring headache, and in the majority of cases are most pronounced early in the morning. They are frequently attended by a slight vertigo and a feeling of faintness, but none of these symptoms is characteristic enough to make the diagnosis of anæmic headache unless the headaches due to eye-strain, to neurasthenia,

or to uræmic conditions can be excluded. The pallor of the conjunctivæ, which may be obscured by a catarrhal condition very frequent in children, and above all the pallor of the gums and of the roof of the mouth, and a deficiency of the hæmoglobin are the signs which help to make the diagnosis of anæmic headaches certain beyond peradventure

The result of treatment will also furnish corroborative evidence. The child should be placed at rest, fatigue should be avoided in every possible way, and its diet should be nutritious to the extreme. It is important not only to give nutritious food, but to exclude everything that is of no decided value to the child's condition. If we allow an excess of fruit or of sweetmeats the child will very naturally look for these rather than for meat, fish, eggs, oysters, which should constitute its main diet, with a slight admixture of farinaceous substances. One to two quarts of milk per day, three or four eggs, a pound or more of meat will be all of far more benefit to the child than any number of drugs. They may help a little, but I am certain that it is only a very little; and if there is any question of the child's power of assimilating these drugs I much prefer to give them up rather than to diminish the amount of food that the child takes. In the treatment of children's diseases the practical necessity of prescribing something often puts physicians at a great disadvantage. The era of rational therapeutics has not yet fully dawned.

These anæmic headaches will be relieved in some instances by the proper use of hydrotherapy. It is a good plan to bathe these children every morning immediately upon rising. Sponge the head and spine first with warm water and then with cold, allowing the water to drip from a little height, so as to get the effect of the impact of cool water. After this they should be briskly dried and allowed at once to dress. In the case of very weak children it would be better to have the child take a little nourishment, say a glass of hot milk, before putting it through this procedure. In addition to this the child should be given regular calisthenic exercises, which help to improve the general condition and to develop the muscles, and to pre-

vent the accumulation of fat, which is not infrequent in children of anæmic disposition.

The meals should be given at regular intervals, and in cases in which the appetite is poor the plan of giving small quantities at frequent intervals, say every two hours, is to be preferred to that of only three meals during the day.

The most useful drug to be employed in the treatment of these headaches is caffeine in doses of one to two grains every hour, or at least three times a day. Black coffee will often answer the same purpose. From the combination of phenacetine and caffeine I have seen very little benefit, and it surely has no advantage over the administration of caffeine alone, and is possessed of the disadvantage that phenacetine acts unfavorably upon the hearts of some children. If iron is to be given let it be administered in the most easily digested form, either in the form of reduced iron, or in one of the many preparations of the peptonates or albuminates of iron that are now in the market. Arsenic can be given in small doses of from three to four drops of the Fowler's solution several times a day. In cases in which a general tonic effect is desired, we can give a palatable combination of iron and quinine with small doses of strychnine, from one-eightieth to one-sixtieth of a grain, three times a day. Why phosphorus should be recommended by some I cannot understand. I am certain that no one can conscientiously assert that he has seen any good effect from its use, and whether given in pill form or in the form of Thomson's solution, the effect in my experience upon cases in which I have known it to be tried has been absolutely nil. The fewer the drugs employed, and the more attention is paid to the hygiene and the diet of the child, the more quickly this anæmic condition will disappear.

2. Headaches due to neurasthenia. These headaches occur so frequently in persons who are at the same time anæmic, that the treatment of one cannot be considered apart from the treatment of the other. In anæmic persons headaches very often do not occur until they have been subjected to some emotional excitement, or have passed through some fatiguing ordeal. These headaches can be recognized as due to some other cause than to anæmia

alone by the presence of other symptoms expressive of the neurasthenic state, such as slight tremor of the tongue and fingers and the exaggeration of the deep reflexes, particularly the knee-jerks; but above all by the very persistent location of these headaches on the crown of the head, and by the description of them as a pressure or a feeling of heat on the top of the head. This description of the headache is as characteristic for children as it is for older patients; and in children emotional conditions, the strain of school work, the rivalry between classmates, is quite as apt to bring about a neurasthenic state and to cause neurasthenic headaches as are the more serious struggles for existence in later life.

The treatment of neurasthenic headaches may be conducted on exactly the same lines as was referred to for those due to anæmia. The hydrotherapeutic procedures and rest are by far the most important, and strychnine and quinine in small doses will be of far more benefit than if anæmia were the only cause; but a cure cannot be easily effected unless all exciting conditions are removed, and to that end the child should be taken from school, and in some instances a change of climate and of home surroundings may be necessary. If this is not sufficient a short rest in bed with forced feeding may be attempted, for the "rest-cure" plan is as efficient in children as it is in adults.

3. Headaches due to transitory hyperæmia. Fluctuations in the blood-supply of the brain sometimes cause severe headaches in children. I have seen many young persons who after some unusual excitement or intense emotion develop severe headaches, associated either with deep flushing, or with marked pallor of the face and with cold extremities. In others, again, persistent headaches come on after an acute exhausting disease, in whom it must be supposed that if there is any transitory hyperæmic condition it is rapidly followed by a lack of blood in the vessels of the brain and its coverings. A chronic hyperæmic condition is probable in children afflicted with heart disease, who are often subject to periods of painful headaches, and a similar explanation must be sought for in young girls just beginning to menstruate, who have severe headaches for

several days preceding and during each menstrual period. While such headaches are often strictly periodic they are very different from ordinary migraine, and can be distinguished from the latter, too, by the late onset of the trouble and by their disappearance as soon as the menstrual function has been properly established.

4. Headaches due to gastric disturbance are a very common occurrence in children of all ages. They are generally frontal, sometimes frontal and vertical. They come on suddenly and persist, as a rule, until the gastric condition has been relieved. These headaches are easily recognized by the symptoms associated with them, viz., fetid breath, coating of the tongue, distress in the epigastric region, flatulency, and constipation or diarrhœa. The diagnosis is easily corroborated by the effects of treatment, a single dose of oil or of calomel being sufficient, as a rule, to dispel them until the next serious error in diet is committed.

5. Genital irritation is referred to by some writers as a cause of severe headaches. Seguin refers to a case of occipital headache, the worst he ever saw, which was cured by circumcision. I have not seen such a case, but I do not wish to doubt their occurrence, and if the physician is convinced of the cause of the disturbance the remedy is close at hand. In older boys, and even in girls, headaches are not infrequently due to self-abuse. The general nervous condition of the patient under these circumstances calls for treatment even more distinctly than do the headaches which are associated with a general restlessness, with pains in the back, with irritability, and in severe cases with a tendency to mental apathy or even dementia. There is every reason to look for this etiological factor in any case of persistent headaches in children between the ages of six and fifteen years. The treatment of the headaches under these circumstances is not an easy matter, and practically they can be cured only by the closest watchfulness on the part of parent or nurse.

6. The headaches associated with ear disease are characterized by intense pain, located either in the mastoid or in the temporal region, and are most frequently present in

the earlier stages of the disease, before the formation of pus. One of the worst cases of this sort that I have ever seen was in a little girl, two years of age, who was unmanageable and showed by her movements that she was in great distress. Every touch on the head was followed by a shrill cry, and the region of the ear was so sensitive that the slightest touch with the finger seemed to cause intense agony. No treatment was of any avail until after a few days the pus was freely discharged through the outer canal, and from that moment every trace of headache seemed to have disappeared. The child's behavior is exactly like that of an adult with acute ear disease; it is true, however, that this special cause of headache is often discovered only by the merest accident. The practitioner and the specialist in ear diseases will meet with these cases so often, particularly in connection with the acute infectious diseases, that they should be on their guard. The cure depends entirely upon prompt treatment of the ear condition.

7. Headaches due to organic disease of the brain or its coverings should be suspected in every case in which the pain is persistent and strictly localized. The pain may vary a little according to the position of the head, but it is present whether the child be sitting up or lying down, and is always elicited by the gentlest percussion of the skull. The headaches constitute a most valuable symptom in the diagnosis of tumors of the brain or of the meninges, and in cases of meningeal inflammation from any and every cause. Thus after minor or greater injuries to the head the onset of intense pain, particularly at the seat of external injury, must lead one to suspect the development of a morbid condition at this point. If due to tumor, the diagnosis can be strengthened by the discovery of other symptoms, which are bound to arise sooner or later. These are vertigo, nausea, and disturbances of vision due to optic neuritis. If a traumatic meningitis is the cause of pain in the head, slight rigidity of the neck, and possibly an inequality and immobility of the pupils, will help to prove the diagnosis.

Among the organic headaches we might also include those due to specific disease; but these headaches, if I may

trust my own experience, are relatively rare in cases of hereditary syphilis or in syphilis acquired at a very early date. They are surely not nearly as constant nor so important a symptom as are the specific headaches of later years. The diagnosis should be made with great reserve, and only if other symptoms are present which point to an active syphilitic process.

8. The headaches which mark the prodromal stage of acute infectious diseases often give rise to very serious errors in diagnosis, and are occasionally suspected to be symptoms of cerebral tumor, or possibly of meningitis. In the prodromal stages of typhoid and scarlatina, and of diphtheria, these headaches are very common indeed; but the possibility of such a cause for headaches should be entertained if they are associated with a general malaise and with slight rise in temperature. There is no need of discussing the treatment of this special form. Unless one chooses to apply cold cloths, or an icebag, or possibly to give small doses of phenacetine, there is nothing else to do but to wait for further developments, and to treat the more serious disease of which the headaches constitute the prodromal stage.

9. Malarial headaches are not observed nearly so often in this climate as in the more southern States, and in those countries in which severe types of malaria are prevalent. Malarial headaches are almost invariably neuritic in character. The supraorbital and infraorbital points are painful, and the entire head may be sensitive to touch. There is a distinct periodicity in the development of these headaches, or if the headaches are continuous there are at least periodic exacerbations.

In making a diagnosis of malarial headaches I follow the practice of examining the spleen and the blood, and not contenting myself with the diagnosis unless I can prove the existence of enlarged spleen, or the presence of the plasmodium in the blood. If such evidence is obtained, a few large doses of quinine—from five to ten grains several times a day, according to the age of the child—will prove the best cure.

10. Uræmic headaches are not as frequent in children as

in the adult. These headaches are generally occipital, and are associated with slight disturbances of vision, with vertigo, nausea, and sometimes with epigastric pain. The condition of the kidneys should be carefully determined, if severe headaches arise during or after any of the acute infectious diseases; above all, in scarlatina and diphtheria, which are known to be followed by renal complications.

11. Other toxic headaches deserve special mention. They are not frequent, but do occur often enough to make it necessary to examine for this possible cause. Among toxic substances lead is most easily productive of severe headaches. The poison is apt to be taken into the system not only with milk that is kept in leaden jars, but with drinking-water passing through leaden pipes, and I have known it to result from chewing-gum, and all sort of vile sweetmeats that are wrapped up in attractive papers, the children licking these papers in order that they may get the full value of their purchase. Some years ago I saw at my clinic a young girl, of about fifteen years, suffering from headaches, and from a typical lead palsy, who had evidently taken the poison into her system in the course of her daily work, which consisted of gluing together the paper in which chewing-gum was wrapped. She was in the habit of putting her tongue to the glue and the paper instead of moistening the former with a sponge.

I have reserved for the last, the consideration of headaches due to eye-strain. Not that I think them the least frequent, but because in my opinion undue importance has been attached to them. The cases that are due to eye-strain are those in which the headaches come on after reading, or in studying. They may persist for some time after the effort is made, but frequently disappear after the effort is relaxed. Serious errors of refraction may be the cause of headaches, and of continuous headaches, even though no effort be made to use the eyes; but I have seen headaches persist so frequently after the fitting of glasses by the most competent oculists that I am firmly convinced that eye-strain is the sole cause of headaches in relatively few instances.

These headaches are located in the frontal region, be-

tween and over the eyebrows (Fig. 51); in some instances with evident eye-strain the headache is referred to the occiput. I am thoroughly in favor of giving every child the benefit of the doubt, and of making a careful examination into the condition of the eyes; but I wish to protest against the excessive enthusiasm of the day which implies that if the slightest error of refraction is discovered in a child the error must be the cause of all ills. What is claimed for headaches is claimed under similar conditions for epilepsy and for chorea.

The above considerations will convince the student that the diagnosis of headaches is no easy matter. To make an accurate, or even plausible, diagnosis of this condition is one of the most difficult tasks in neurology. It can be done properly only by a careful consideration of the general condition of the patient, of the symptoms associated with the headaches, and of the patient's health before and after the headache has been developed.

MIGRAINE.

Among the neuroses of early youth few are more troublesome or more interesting than migraine, or "sick headache." While the disease does not, as a rule, attain its full development until the age of puberty or later, it begins so often in the earlier years of life that it belongs very properly to the special subjects of this book. The symptoms of the disease as it occurs in children are so very much like the adult form that in describing one we picture the other.

Hemicrania (megrim), or sick headache, is characterized by occasional attacks of intense headaches, frequently unilateral, which are associated with a feeling of nausea, or with vomiting. The unilateral headache is by far the more important symptom of the two, for in many cases the characteristic headache is present for months and years without nausea ever being associated with it, though I must record cases from my own experience in which periodic attacks of nausea have occurred in children without any headaches. Such attacks I have interpreted to be the equivalent of or-

dinary migraine. The correctness of this view has been proved by the occasional occurrence of attacks in which both nausea and headaches were present, and by the fact that the ordinary treatment for migraine and no other helped to dispel these peculiar gastric attacks. Barring such occasional cases, it is better for us to consider the typical attack in which neither headache nor nausea is wanting.

In this disease we find a number of very important symptoms associated with headache. Peculiar visual disturbances constitute a prominent feature of the disease; these have been variously described by many sufferers, and amount, as a rule, to a temporary and partial loss of sight during the attack; or there may be every possible form of visual disturbance, from simple balls of fire to distinct figures, which appear as regularly in the attacks of migraine as similar phenomena do in attacks of epilepsy. In many cases a simple dark spot is observed; in others, flashes of lightning that surprise the patient are the first symptoms of a full-fledged attack of migraine. A young patient of mine would regularly see a bright zigzag line, which she compared to a distant staircase; as soon as she seemed actually to approach the stairs intense headaches would set in, and she would feel dizzy, but there was never loss of consciousness, and nothing resembling an epileptic attack. The patient would then pass through a typical attack of migraine, which would last for several hours, and during this entire time would exhibit very marked photophobia. In some cases other special senses are affected. Tinnitus is much more frequent, on the whole, than any other form of sensory disturbance excepting those of vision noted above.

Disturbances of sensation in the limbs are a frequent accompaniment of migraine. These sensations generally take the form of tingling, of pins and needles, or of burning sensations. In some instances there is a general numbness, very much like the numbness that precedes an attack of epilepsy; the sensations, however, last very much longer than the sensory aura would, and can, of course, be distinguished from the latter by the entire absence of any typical clonic movements. Motor symptoms are not present as a

rule. As there is a close association, at times, between migraine and epilepsy it is possible that we may see cases every now and then in which the auræ resemble very much those that precede an attack of migraine, but the clonic movements are characteristic of epilepsy.

Temporary aphasia I have met with as an accompaniment of an attack of migraine. I recall the case of a young girl, ten years old, who had inherited migraine from her mother, and who greatly alarmed her parents by the sudden development of aphasia in connection with an attack of migraine. The girl when spoken to was able to mumble a few words indistinctly, but could not find the word she wished to say. She was in intense pain and extremely irritable, but, after a good night's rest, the headache had disappeared and with it the aphasia. This aphasia is associated with right-sided hemiplegia in right-handed persons. The occurrences during these attacks of migraine adhere closely to the physiological laws of the cortex, and we may anticipate what we have to say upon the pathology of the subject to the extent of implying that the sequence of symptoms evidently proves that the entire motor district of the brain must be affected by the temporary defect in its blood-supply.

Vertigo is another symptom that is associated with the headaches, at times preceding it, at other times following it. The vertigo is not, as a rule, as marked as it is in Menière's disease or in some organic diseases of the brain, but it is quite sufficient to make the patient unsteady on his feet, and to give rise to a great deal of discomfort.

The aspect of the patient varies considerably during the attack. In some there is distinct pallor of the face and a feeling of coldness in the extremities. The eyeball may seem a trifle retracted, the vessels of the conjunctiva may be engorged, and the pupils may be contracted. In other cases the face may be extremely flushed, the pupils dilated rather than contracted, and the ear distinctly reddened. These two distinct conditions are often present in one and the same attack, and, in rare instances, the one-half of the face may present pallor with its associated symptoms, whereas the other half presents a flushed condition with

the symptoms that go with it. The latter symptoms are evidently of the paretic order and the former of the spastic, and both are evidences of a change in the sympathetic nerves. Sweating of one side of the face has also been observed, as well as retardation of the pulse during the paroxysms. On examination during an attack, I have found the various points of the trigeminal nerve quite as sensitive as in the milder cases of trigeminal neuralgia. In some patients—and this is particularly true of children—the entire face and head is so sensitive that the patient can scarcely bear to rest the head on a pillow, and I have known one patient who insisted on "walking off" the headache rather than to rest the head against anything during the paroxysm.

Gowers refers to a transitory pyrexia during an attack of migraine in children. I have often taken the temperature during attacks, impelled to do so by a flushed condition of the face, but have never found any elevation above 100° F. If a higher temperature is present I should be disposed to look for some other condition, say some gastric disturbance, as a possible cause both of the migraine and of the fever.

ETIOLOGY.—The hereditary predisposition to migraine is too manifest to be denied for a single moment. Not only do children of mothers who are thus affected inherit the disease, but it also occurs in the progeny of persons afflicted with other grave forms of functional nervous disturbance. Among these epilepsy, hysteria, and hystero-epilepsy are the most potent sources. The disease usually sets in in the earlier years of life. Some begin between the fifth and tenth years, a fair number of the cases between ten and twenty years, but the majority set in between twenty and thirty years.

The female sex is evidently more disposed to the disease than males.

The first manifestations of migraine are generally excited by emotional disturbances, by overwork, or worriment. In not a few instances I have known the rivalry among classmates to have been the final cause of the development of migraine; in others, some severe gastric disturbance has

been the starting-point of the entire trouble, but it should be remembered that, if a predisposition did not exist, the derangement of the stomach might have produced a temporary headache, but would not have started a series of attacks of migraine. After the first attack an interval of some weeks, or months, may intervene before a second seizure takes place; but some form of periodicity is soon established in the majority of the cases, and in many, even in young children, the attacks are apt to return at stated intervals—every two, every four weeks, or every second or third month. It is with migraine very much as it is with epilepsy, that the slightest disturbance of the physical organism is sufficient to develop an attack. It is, therefore, of the greatest importance in migraine, as in epilepsy, that the most careful attention should be paid to the general hygienic and dietetic management of the child.

Pathology.—We cannot expect to demonstrate actual changes in the brain, or in any other part of the central nervous system in a person afflicted with migraine, for persons so afflicted rarely die after a paroxysm, and the attack of migraine evidently represents a transitory change; but since the anatomical basis is wanting, the opportunity for theoretical speculations regarding the pathology of the disease is all the greater, and has been improved by numerous writers. There can hardly be a doubt that changes in the blood-supply of the brain or its coverings are primarily responsible for the symptoms of migraine. There is also reason to believe that the sympathetic nervous system is largely involved in this disease. According to the varying conditions present many neurologists are inclined to suppose that in some instances we have an angio-spastic condition, in others an angio-paralytic state, and if we wish to imply our belief in the *rôle* played by the sympathetic nerves we can speak of "sympathetico-tonic" or "sympathetico-paralytic" forms. The behavior of the superficial blood-vessels in the two forms of migraine lends reasonable coloring to this view of the vasomotor origin of migraine.

Some, not satisfied with this vascular theory, argue that there must be some inherent alteration in the nerve-cells of the brain, but no sufficient reason is given why, if such de-

rangement exists, there should be such violent periodic exacerbations of all the symptoms. To say that the symptoms are due to a "nerve storm" is merely substituting a vague term for a vague conception; yet we must acknowledge that a peculiarity in the structure or in the function of the cortical cells may be present in these cases, and that changes in the vasomotor apparatus are sufficient to produce the phenomena of migraine in a person whose nervous system is thus altered, whereas the same vasomotor changes would be entirely insufficient to produce any such symptoms in persons whose brain-cells are altogether normal. It seems to me, therefore, that we must rely upon these two causes for an explanation of the disease; surely one cause alone would not be sufficient to explain all the phenomena. We are constantly losing sight of the co-operation of forces and of causes that produce disease, and are hampered so frequently by the supposition that we must make out a single cause or none.

The disturbances of vision, as well as the temporary aphasia occurring in some cases, prove conclusively that the cortical centres are involved in the disease. The visions of migraine can be regarded as a symptom of irritation, the hemianopsia as a symptom pointing to temporary paralysis of the functions of the visual centres. Whether a derangement of the cells or some other change in the constituent parts in these centres is responsible for the loss of function cannot yet be proved or denied.

The relation of migraine to epilepsy makes it also more probable that some primary peculiarity of the cortical cells is responsible for this painful affection. The resemblance is so close, and the sequence of the two diseases so striking, that the two forms of disease may possibly represent a different degree of affection of the cortical structure.

As for the headaches of migraine, they can be best explained, it seems to me, on the supposition of a marked distention of the blood-vessels of the coverings of the brain; and the fact that various points in the face are as painful as they are in typical trigeminal neuralgia, would lead to the inference that the trigeminal nerve, as it passes from the brain outward, is affected by this general hyperæmic con-

dition. The nausea and vomiting are an expression of general cerebral disturbances, such as is seen in cases of gross disease anywhere in the brain, particularly in the lower centres in the pons and medulla, and such as we sometimes find in cases of simple cerebral shock. I cannot see sufficient reason to connect these symptoms directly with an affection of the sympathetic nerve.

Within the last few years much has been made of ocular insufficiencies as a possible cause of migraine. This matter has been much overdone, thanks to the labors of Dr. Stevens and others. As a neurologist I could pass over the rather elaborate discussion that has been held on this subject, were it not for the fact that so high an authority as Dr. Seguin has given his approval to this special doctrine in his lectures on the treatment of neuroses (*New York Medical Journal*, 1890). Dr. Seguin states that he has not met with a case of migraine in a person with normal eyes, although he has been told of two or three by oculists of good repute. He implies, furthermore, that if the matter has been overlooked in the majority of cases of migraine, it is because the ocular examination has not been a thorough one. He argues, furthermore, that the ocular origin of migraine is made probable by the remarkable fact that in many persons of both sexes the attacks diminish and then cease between the ages of forty and fifty years, at the time the power of accommodation becomes exhausted, and a large part of the unconscious strain which has been going on from early youth is removed. Seguin implies still further that migraine is hereditary, chiefly because ocular defects are hereditary. The fallacy of these arguments seems to me to be evident enough from the experience many of us have had, that the eyes are normal according to the examination of competent oculists in many children who have migraine, and that the attacks continue long after the ocular difficulty has been corrected by glasses. We are willing to concede that these ocular difficulties have an important bearing upon headaches in general, and may influence the occurrence of attacks in those who have inherited migraine; but far stronger evidence will have to be advanced before it will be safe to adopt Seguin's conclusions, and some other

sufficient reason will have to be given for the action of mydriatics, such as belladonna, atropine, hyoscyamia, and cannabis indica, than the effect these have upon the accommodative effort. In my own experience with the drugs mentioned, cannabis indica is the only one which has any decided effect upon the course of migraine or upon single attacks.

DIAGNOSIS.—The diagnosis of migraine is easily made. The early onset of the disease, the periodic attacks, and the character of the headaches will, as a rule, leave little room for doubt. It is only in those cases in which a distinct sensory aura exists that a confusion with epilepsy might arise, but the preservation of consciousness and the entire absence of clonic movements, and the duration of the single attack, will help to distinguish migraine from epilepsy. It may be a little more difficult at times to distinguish between migraine and headaches due to some other cause, but this difficulty will cease as soon as the periodicity in the occurrence of the headaches has been noted, and every other form of headache has been ruled out by the method of exclusion. I have found some little difficulty also, particularly in young children, to distinguish between migraine and ordinary trigeminal neuralgia, for owing to the sensitiveness of younger patients it is not so easy to determine whether there are distinct painful points, or whether the whole face is painful, merely as a result of the fear of examination; but the long and free interval between attacks, together with the periodic occurrence of the headaches in migraine, will serve to distinguish this form from typical trigeminal neuralgia.*

PROGNOSIS.—The prognosis of migraine is entirely favorable as regards life, but not so promising with reference to the cure of the disease. It is a matter of common experience that a person who has inherited migraine will be subject to the disease for a long number of years, until it begins to disappear with age. It is a satisfaction, however, to be able to assure the patient that under proper treatment the attacks may be either inhibited or their severity may be diminished. The only grave feature about the disease is

* Trigeminal neuralgia is a rare affection in children.

the possible development of migraine into epilepsy, but even this is an infrequent occurrence.

TREATMENT.—In the treatment of migraine two distinct objects must be kept in view. First—the cure, if possible, of the disease; secondly—the amelioration of the attacks.

The disease often defies the skill of the most experienced practitioner. As in epilepsy, no effort should be spared to accustom the patient to regular hygienic and dietetic habits. It is important that the child thus afflicted should sleep in a well-ventilated room; that it should have regular and moderate physical exercise, a point of the greatest importance in those who have inherited gouty or lithæmic tendencies. The child should be relieved also, as far as possible, of all mental and physical strain. While the disease is at its worst the patient should be kept from school, and all close application to study should be avoided. If it can be shown that the effort of reading or of studying helps to bring on an attack, even study at home and reading may have to be prohibited. As a matter of fact, however, a little mental occupation is often to be preferred to idleness, which permits the child to wait for the onset of an attack and often induces distinct hypochondriacal tendencies. My own experience has been very largely in favor of moderate employment of mind, which helps to prove to the child that its lot is not a worse one than that of other children, and helps also to avoid the idea of invalidism which is so frequent in children, even in those who do not for one reason or another care to simulate disease.

If the child is anæmic, or if it exhibits scrofulous tendencies, iron in any of its various forms, arsenic and quinine in tonic doses, as well as cod-liver oil, are indicated. Phosphorus is of so little use that we can disregard it altogether.

A single attack can be relieved best by putting the child in bed during the period of the attack, and in a darkened room. On the whole mere rest in bed is as important a factor as any in the treatment of the attack. During this time the child should be placed on a mild diet; an excess of liquids as well as of nitrogeneous food should be avoided. Small amounts of meat, fish, and eggs, and a moderate

amount of milk, will answer the purpose better than if the patient is placed altogether upon nitrogeneous or upon an excess of farinaceous food.

It is important to regulate the bowels thoroughly, and if the kidneys are not very active to increase the quantity of urine passed. To this end some mild alkaline water, such as Vichy or Seltzer, can be safely recommended, either alone or in conjunction with milk.

Innumerable drugs have from time to time been warmly recommended. Some years ago antipyrine in doses of five to ten grains, according to the age of the patient, was quite in vogue; but I do not consider it promising enough to urge its use in the face of the well-known risks attending its exhibition. I am certain, too, that phenacetine in five to ten-grain doses has little or no effect. I have seen some good results from the combined action of the phenacetine and caffeine in one- to three-grain doses, but I am certain that the combination is not in any way superior to the use of caffeine alone. Caffeine is beyond a doubt one of the most serviceable drugs to be employed. I am in the habit of giving it to children under fifteen years of age in one-grain pills or powders, which are to be repeated every twenty minutes until distinct relief is felt; and after the first relief has been procured the drug should be continued in the same dose at longer intervals, first every two, then every three hours, and later on, for a period of about a week after the onset of the attack, it can be given safely in small doses several times a day, according to the condition of the child. Caffeine can be combined with iron, with quinine, or with arsenious acid. I have also had excellent results from a combination of caffeine and cannabis indica. The various preparations of the latter are so unreliable that it is as well to use Herring's extract (gr. $\frac{1}{10}-\frac{1}{6}$). A caution is necessary, however, as regards the use of cannabis indica, for some patients are peculiarly intolerant toward this drug, and I have seen distinct symptoms of poisoning from small doses repeatedly employed. While the drug is not in reality a dangerous one, the symptoms due to the administration of large doses are so disagreeable and are so apt to alarm the patient and the family that great care should

be exercised in the use of the drug; but if the fact of tolerance on the part of the patient has once been established no trouble is apt to ensue during subsequent trials. Seguin thinks that the drug exerts a sedative, even paralyzing, influence upon the third cranial nerve and its attached muscles, including the ciliary, but the drug has as good an effect in cases in which the ocular apparatus has been determined to be entirely normal.

The nitrite of amyl has been suggested on physiological grounds as a proper remedy, particularly in those cases in which there is marked pallor of the face. We might also expect it to act favorably, as it does in epilepsy, in those cases of migraine in which there is a distint sensory aura. The bromides and chloral help to allay the nervous excitement and to induce sleep, as do also sulfonal, trional, veronal, and chloralamid in four to fifteen-grain doses; but I cannot regard them as in any sense specific remedies in migraine. Nitro-glycerine (one drop of the one per cent. alcoholic solution) has occasionally helped to diminish the severity of an attack. In this form it is not a dangerous drug, and can be safely administered under the supervision of a physician, of a nurse, or of an intelligent mother. The use of tea and coffee in moderate quantities is strongly to be recommended; I have known children suffering from migraine to do well on black coffee who would vomit everything else that was given. I feel that I owe much to Dr. Seguin's suggestion that black coffee, without sugar and without milk, is easily digested, and that the general prejudice regarding this beverage is due to the milk and sugar that have been invariably added, and that have given rise to fermentative processes in the gastro-intestinal tract.

As for the correction of the ocular defects, I do not wish to oppose sensible treatment by competent oculists, but I do oppose treatment based upon extreme theories. I would have the eye condition corrected on the principle that in migraine, as in epilepsy, every physical defect should be remedied; but I must insist that such slight errors in refraction and in accommodation are not the cause of this neurosis.

CHAPTER X.

THE DISORDERS OF SLEEP.

THE healthy new-born child sleeps during the entire day and night, except when it is being nursed or dressed. During the first month the infant is awake only three or four hours out of the twenty-four, falling soundly asleep immediately after nursing. By the end of the sixth month fifteen hours' sleep per day is a fair average, which is gradually decreased until at the age of one year the healthy child sleeps about twelve out of the twenty-four hours. From one year to four years an average of ten hours' sleep should be maintained, and up to the age of ten or twelve years the health of the child demands that it shall sleep quietly for at least nine hours in the day. If there is a marked departure from these averages, the child is either in pain or some of its functions are not entirely normal. In the acute fevers or in any febrile condition children are alternately drowsy and wakeful. Disturbances of digestion are also a frequent cause of restless sleep or of prolonged wakefulness.

In addition to these special conditions, others cause insomnia in children as in adults. Poorly ventilated and overheated rooms often cause restlessness, and for this reason it is important that a window should be kept open in every bedroom during the entire night, and that the child should be covered as lightly as possible, according to varying atmospheric conditions. If a child is sleepless in the absence of unfavorable conditions, we must seek some other cause for the insomnia. In many cases the mother or nurse has not been careful to engender the proper habits of sleep. Infants that are much pampered, that are taken up as soon as they utter the first cry, or that are wilfully disturbed by parents or nurse, soon become poor sleepers. From the first week of life the child should be laid down quietly and

not rocked to sleep, and should not be disturbed except for good reasons. If the room is darkened a quiet sleep will come on very naturally. As the child grows older it is well not to excite it just before the hour at which it is expected to fall asleep. With children up to the age of eight years the evening meal should be a light one, and their brains should not be disturbed by exciting stories. This should be observed more particularly in the case of children who exhibit nervous tendencies, or of those who have passed through exhausting diseases.

The normal child does not fall asleep unless its hunger has been appeased. Insufficient nourishment is, therefore, to be suspected in cases of persistent wakefulness in young infants. With the change of nurse, or after weaning, a restless sleeper will often develop normal habits of sleep. If insomnia continues without apparent cause, we must suspect some graver trouble. In the earlier stages of tubercular meningitis, in tumor, and in other painful diseases of the brain, sleeplessness is often one of the earliest symptoms.

If the cause of insomnia cannot be easily made out, it is well to fall back upon faulty methods of training. Not very long since a child of five years was brought to me by its mother, who stated that it would not fall asleep if it awoke after midnight. The mother claimed to be ignorant of any mistake that she had made in the training of the child, but on closer questioning I heard that she had been in the habit, whenever her husband was away, of taking the child out of its bed and putting it in her own bed. After she had done this a number of times the child woke up regularly at midnight and would cry until the mother took it up. The mother, a very nervous woman, excited the child by her caresses and her despair over its not sleeping, and thus made matters worse and worse. The child had been sleeping very poorly for several weeks before I was consulted. The cure was a very simple one. The child was placed under the care of a sensible trained nurse, who would not yield to its solicitations, and after a few nights of restless sleep it began to develop proper habits and soon slept an average of ten hours.

The prognosis is invariably favorable unless the condi-

tion is due to some grave disorder. It is most important to insist upon absolute regularity in putting the child to bed at a definite hour and keeping it there unless it is necessary to take it up. If this should prove insufficient a warm bath at bedtime will be conducive to sleep, and small doses of bromide or chloral will be useful to engender the habit. Sulfonal or trional, in five- to ten-grain doses, veronal in three-grain doses, may be substituted for the bromide or chloral in children who are anæmic or poorly nourished. All hypnotics should be withdrawn as soon as possible, for there is great danger of accustoming the child to these drugs. In some cases the mere regulation of the meals, and the avoidance of all indigestible articles of diet, will be sufficient to restore sleep. In children who are impressionable it is of the greatest importance to avoid telling them stories or even engaging them in play in the latter part of the day.

Pavor Nocturnus, or night fear, is a condition which is often troublesome both to the parents and the physician. Children affected with this disturbance wake up a few hours after they have entered into a sound sleep, are possessed as a rule by great fear, fail to make out their surroundings, and act as though in a temporarily dazed condition. They do not understand the soothing words of parent or physician, and often continue in a state of excitement until they return to full consciousness or are overcome by sleep.

There seems to be no good reason to give any other interpretation of this condition than that the child has had a horrible dream, and that the substance of the dream is continued in the half-waking state, or that the child is possessed by fear, which is the natural result of the dream and cannot shake off the fear until it fully realizes, if old enough, that its experiences have been unreal.

This condition may occur in any child, but it is decidedly more frequent in children who have shown previous nervous symptoms, or in children of highly neurotic parents. It is not infrequent in those who are pressed by school duties, and who go to bed with a fear that their lessons have not been properly done for the next day. This knowledge is sufficient to prevent sound sleep, and during the hours of

restlessness vivid dreams are very apt to occur. It was formerly supposed that late and indigestible meals were the chief cause of these noctural disturbances. Inasmuch as any indigestion may give rise to restless sleep, this factor may have to be taken into account; but over-work, over-excitement, the reading of horrible or fascinating tales just before bedtime, constitute a much more important etiological factor.

These noctural attacks often occur a single time, but with most children are frequently repeated during the earlier years of life, say between the ages of three and eight years. With the growth of the child and the improvement of its nervous system the attacks cease.

The prognosis of this condition is entirely favorable; though I can recall one case of a girl, aged six years, the child of an hysterical mother, in whom the fright connected with one of these nocturnal attacks was the cause, or at least the first beginning, of a severe chorea, which lasted for many months. In another child, about nine years of age, who had had attacks of epilepsy ever since early childhood, and who was particularly subject to nocturnal attacks of epilepsy, the epileptic attacks ceased upon bromide treatment, but every now and then the child would have a very marked spell of pavor nocturnus, which would disturb her almost as much as an epileptic seizure would.

Conditions similar to those of pavor nocturnus occur at times during the day. A little patient of mine, aged five years, would, in a very unaccountable manner, while walking with her mother on the street, suddenly hide her face in the folds of the mother's dress, and bury her head for some time, in order (as she explained later on), not to see the horrible black things coming toward her. This child showed no other symptom of a nervous disposition, but was for a time in great dread of the recurrence of these experiences. That they were the result of visual hallucinations during the waking hours, I can hardly doubt. They disappeared entirely under sedative treatment, which was given carefully for some months.

In regard to treatment, the most important point is to exercise every possible precaution in order that unnecessary

nervous excitement, late in the day, may be avoided. I prohibit all school-work after five in the evening; do not allow the child to read or be read to after this hour, and insist on a very light meal in the evening. After this meal the child is to remain awake for at least an hour, and may indulge in some simple game, or is allowed to frolic about in a quiet manner. Small doses of the bromides, from five to ten grains, according to the age of the child, about an hour before bedtime will be the best remedy. If necessary, small doses of chloral may be given with the bromides. If the attacks return very frequently I have found it more satisfactory to give five- or ten-grain doses of chloralamid, or of trional, every night, or every second or third night, until all excitement has subsided.

Enuresis Nocturna is another very common form of disturbed sleep. Children who have been well trained, and who have learned to observe all the habits of cleanliness during the day, are frequently disturbed by involuntary micturition during the night. This is most apt to occur during the earlier hours of sleep, when sleep is most profound. The soundness of sleep alone may be a sufficient cause for this phenomenon, inasmuch as children do not perceive in sleep those symptoms of vesical irritation which prompt them to evacuate the bladder during waking hours, and during light sleep. In others, and possibly in the majority of cases, it is due to indifference on the part of the child, or to an aversion to allow its sleep to be interrupted. In some few cases enuresis is the result of frightful dreams, the sudden impulse to evacuate the bladder accompanying such a dream as it would any emotional condition during wakefulness. If due to this latter cause the enuresis is not frequently repeated. Enuresis has been observed to be a symptom of nocturnal epilepsy. If so, it occurs at rare intervals; the child is entirely unconscious of it, and often shows some other symptom, such as drowsiness and headache on the following morning.

The condition is a very stubborn one, and all possible methods of treatment have been suggested. I am convinced that the cure of the condition is dependent chiefly upon careful training. The necessity of evacuating the

bladder should be impressed upon children after the age of two years. The child should be made to void urine immediately before going to bed, and should be aroused one and a half to two hours after it has fallen asleep, and should be induced to pass water again. By giving relatively few liquids after four or five o'clock in the afternoon, I have found that the tendency to enuresis is often checked.

If these simple measures are not sufficient, a few drops of the tincture of belladonna, given at bedtime, will be useful, and if the condition is due to frightful dreams, or to cerebral excitement continued during sleep, small doses of bromides may be given. Many medical men have seriously suggested that corporal punishment applied to the nates is the only efficient remedy, and some have gone so far as to suggest that the cutaneous hyperæmia caused by such punishment explains the relief afforded. This explanation is not satisfactory ; but whatever the mode of action may be, it is very certain that such punishment need not be inflicted in the majority of cases, and that the desired end can be attained by other methods of training.

SOMNAMBULISM is by no means rare in children, and occurs most frequently in those who are of a nervous temperament, or who have passed through some severe excitement. I have not known them to perform any of the marvellous tricks which are generally accredited to somnambulists, such as climbing out of windows and on the roof, and the like ; but they are apt enough to walk through the house, from one room to another, and to go through a number of purposive actions without being at all conscious of what they are doing. Nightmare is evidently closely related to this condition of somnambulism, and is generally associated with horrible dreams, from which the child awakens badly frightened, but is quieted much more rapidly than in the condition which is described as pavor nocturnus. Nightmare and somnambulism occur, as a rule, at rare intervals and can generally be traced to some emotional excitement, or to some acute indigestion. The treatment of these conditions is exactly the same as that of night-terror.

CHAPTER XI.

VASOMOTOR AND TROPHO-NEUROSES.

In this chapter a brief description will be given of a number of rare and peculiar diseases, which must for the present be classified under the rather vague title of vasomotor and tropho-neuroses. Grouping them together under one head is simply a matter of convenience.

EXOPHTHALMIC GOITRE.

This disease, which is generally termed Graves's disease by the English writers, and Basedow's disease by the Germans, occurs chiefly in adult life, yet a sufficient number of cases begin during childhood to make it incumbent upon us to give a brief description of the disorder.

The cardinal symptoms are:

1. Excessive cardiac action. This is by far the most constant symptom of all, and is often present in the earlier stages of the disease when the other two cardinal symptoms —swelling of the thyroid and the protrusion of the eyes— are not yet in evidence. The pulse-rate may vary between 90 and 200. In the later stages of the disease the left ventricle, and rarely the right ventricle, may become hypertrophied and dilated. The heart-sounds are clear but unusually loud. All the arteries pulsate very distinctly, so that the pulse of the abdominal aorta may possibly be observed through the abdominal walls, and the pulse in the carotids may be so strong as to cause distinct pulse-like vibrations of the entire head. The pulse may even be noticeable in the retinal arteries, and a further disturbance of the entire vascular system may be noted by a dilatation of the capillaries in the skin, by the presence of distinct *tâches cerebrales*, and a dilatation of all the larger superficial veins. If

venous stasis is marked, œdema may occur in various parts of the body. It is also owing to the increased tension that hemorrhages are frequent from the nose, and even in the stomach and bowels. Cerebral hemorrhages have also been described in Graves's disease. A particularly characteristic symptom, which is found in exophthalmic goitre, and not in any form of enlargement of the thyroid, is the whirring felt if the finger is placed upon the goitre.

2. The second cardinal symptom is the enlargement of the thyroid gland. It is present in the vast majority of the cases, though a number of authors have taken pains to describe cases of this vasomotor disease in which the thyroid was not enlarged; but the enlargement, as a rule, comes on some time after the tachycardia. The gland, after it has once become enlarged, increases rapidly, and may give rise to a great deal of inconvenience. The enlarged gland, or struma, is extremely vascular, and the tissue of the gland is not only hyperæmic but very soon enters into a state of hyperplasia with a tendency to a fibrinous degeneration. In keeping with these changes the goitre may in the beginning be soft and yielding, later on it will become hard to the touch. The enlargement of the thyroid is, as a rule, bilateral, but generally asymmetrical. The right half is more frequently enlarged than the left half.

3. The third cardinal symptom is exophthalmus, or protrusion of the eyes. This is the least constant of the three symptoms. If present it is generally bilateral, but in some instances one eye only may be affected, or the protrusion of one may be very much greater than that of the other eye. The causes of this protrusion have been in dispute, but it is more than probable that the interference with the venous current, as well as the arterial congestion, are the prime causes of this protrusion. The great variability of this symptom during life, its disappearance almost immediately after death, and the pulsation of the vessels in the fundus, lend support to this view. It can hardly be doubted, however, that in some cases the development of fat and of cellular tissue in the retro-bulbar space may make the protrusion still more marked.

Vision is not affected in spite of the exophthalmus, and

the only changes observed were those recorded by Kast and Willbrand, who have noted a limitation of the field of vision in this disease without any evidence of hysteria. The pupils may be unequal, but react promptly. Ulcerations of the cornea, occurring in consequence of insufficient protection of the bulb, have been noted; as well as some interference with the lachrymal secretion.

Among the accessory symptoms those relating to the movements of the eyes are the most important. Graefe's symptom refers to defective movements of the upper eyelid on vision downward. The upper lid does not follow promptly the downward movement of the bulb, and the conjunctiva remains visible between the pupil and the margin of the lid. This symptom evidently bears some relation to the phenomenon described by Stellwag consisting of a dilatation of the palpebral fissure. Graefe's symptom, as far as can be ascertained, is caused neither by the protrusion of the eyes nor by a spasm of the levator or a paresis of the orbicularis, but is explained most readily as the result of Stellwag's phenomenon. Moebius has described another symptom implying a deficiency in the power of accommodation. There is defective convergence of the axes of the eyes. These three symptoms are not absolutely pathognomonic of Basedow's disease, as each one has been described in connection with other neuroses; but if they are associated with either one of the cardinal symptoms they help to corroborate the diagnosis of Basedow's disease.

The next most frequent, and perhaps most important, symptom is a tendency to profuse diarrhœa. This is entirely independent of any gastro-intestinal disease, often resists treatment, and adds greatly to the patient's discomfort and to the general depreciation of his health. Gastric crises have been described in some cases, and frequent vomiting is not an unusual symptom. Most patients exhibit considerable dyspnœa, resembling true asthma, and Louise Bryson has referred to defective expansion of the chest on inspiration. Glycosuria and albuminuria have been reported; the former, according to the investigations of Chvostek, is of the alimentary order. In some few instances, however, diabetes mellitus has occurred as a complication

of Basedow's disease. This association is not a strange one, since both diseases are often developed after severe emotional excitement. Of the disturbances in menstruation we need take no account, as we are, after all, concerned with the disease as it occurs in children.

Trophic and vasomotor disturbances of the skin are frequent. Pigmentation, leukoderma, loss of hair, premature grayness of hair, and even scleroderma, as well as herpes, have been observed. A very remarkable symptom of Graves's disease is a tendency to profuse sweating. This may be universal or partial, sometimes strictly unilateral. This tendency must be held to account for the remarkable diminution in electrical resistance which has been observed in the skin of patients suffering from this disease. This fact was first insisted upon by Vigouroux, and later by Eulenburg. Kahler and others have shown that this reduction in resistance to the galvanic current is due to the fact that the relative minimum of resistance is reached much more easily in patients suffering from Graves's disease than in healthy individuals. Kahler has also shown that this diminished resistance is not due to an hyperæmia, as was supposed by some, but it is due to excessive moisture of the skin. A faint tremor is frequently observed, and by some is considered to be one of the cardinal symptoms, but it is not as constant as the three cardinal symptoms we have mentioned. The tremor is rhythmical and consists of eight or nine vibrations per second. Choreiform tremor occasionally occurs, and true chorea and epilepsy are complications met with in a certain number of cases. These complications are in all probability purely accidental, and cannot readily be explained on the supposition that the vasomotor changes in Basedow's disease are the cause of the chorea and epilepsy.

In addition to the preceding symptoms mental changes deserve some notice, and among these we may note irritability of temper, sudden changes of temperament, and the occasional occurrence of mania or melancholy. Some of these psychic changes are developed in the earlier stages of the disease, others are later, and possibly accidental complications. If we add anæmia, albuminuria, and a general

cachexia, we have completed the list of the symptoms typical of Graves's disease.

The morbid anatomy of Graves's disease has been the subject of much discussion. The cervical sympathetic has been given a very important part. A number of authors have reported actual changes in the cervical ganglia as the result of connective-tissue proliferation, and others have reported atrophy and degeneration of the nerve-elements; but since Hale White has proven that similar changes occur in persons who have not exhibited the symptoms of Graves's disease, and that in persons who have died from this disease such changes have not been found, the force of all these findings has been much diminished. The same may be said of hemorrhages into the fourth ventricle, or atrophy of the corpus restiforme, to which Mendel and Leube have attached some importance. The heart has naturally exhibited hypertrophy or dilatation, insufficiency of the mitral valve, and symptoms of mild endocarditis, but these changes are clearly secondary and do not at least hold any causal relation to the disease.

The theories regarding the pathology of the disease have been still more conflicting. Basedow supposed the disease due to chlorotic changes in the blood; but this explanation is evidently insufficient, as the disease frequently affects persons in robust health who are far from anæmic. The close resemblance between the symptoms following upon section of the cervical sympathetic and those of Graves's disease has led many writers to attribute exophthalmic goitre to disease of the sympathetic! Section of the sympathetic produces irritation, and the symptoms following the experiment upon animals are clearly the result of irritation; but we cannot suppose any condition of irritation to last for years, and must therefore seek some other explanation. Friedreich was inclined to the theory that dilatation of the vessels is the result of paralysis of the sympathetic; that dilatation of the coronary arteries caused tachycardia, and that this increased flow of blood to the cardiac muscle produced an increased activity of the heart, and that all the other symptoms of Graves's disease, the exophthalmus and the thyroid swelling, were the result of the arterial hyper-

æmia. Granting that the cardinal symptoms can be explained in this way, the many accessory symptoms cannot be accounted for on this theory. Other authors have insisted on the bulbar origin of Graves's disease, and have supposed that a lesion involving the nuclei of the vagus was sufficient to account for all the symptoms, and some post-mortem findings have given support to this view.

A number of authors, more especially Moebius, have insisted on the probability of the toxic origin of the disease, and have supposed that by the increased action of the thyroid gland a toxine is produced which is primarily responsible for all the symptoms. In a recent and very able review of all the conflicting theories (hæmatogenic, neurogenic and thyreogenic), Eulenburg (Die Deutsche Klinik, 1904) concludes that there must be qualitative as well as quantitative changes in the secretion of the thyroid gland to account for the symptoms of Basedow's disease. These quantitative changes, Eulenburg supposes, are due to an abnormal nerve influence, the exact nature of which is still to be determined.

The course of the disease is, as a rule, extremely chronic. The onset may be very sudden, and even violent, but after a while the symptoms subside in severity and remain constant for a long period of years; but the prognosis is not necessarily as grave as it is generally supposed to be. I have myself seen a number of cases of very marked improvement and of total disappearance of all the symptoms. If death ensues it is, as a rule, due to some complicating disease, or to extreme exhaustion from diarrhœa, or from cardiac weakness, or possibly from acute mania.

TREATMENT.—Absolute rest and careful feeding are the most important measures. The recoveries which I have seen have been in patients who have been placed on the rest-cure. The entire freedom from excitement, and regular feeding have tended to lessen all the nervous symptoms and have influenced the diarrhœas which are often the most exasperating and exhausting symptom. The tachycardia is remarkably lessened and the dyspnoea is naturally benefited by rest in bed. The diet should be simple. It is well to avoid stimulants and excitants as well as carbonated waters. The excessive dilatation of the stomach, resulting from the

last-named, acts unfavorably upon the heart. Hydrotherapeutic procedures, consisting of cold baths followed by massage, of douches and drip sheets, have been recommended by many. Mineral baths may have some effect upon the constitutional symptoms and thus favorably influence the disease. Electricity has been warmly recommended by many, with special reference to the part played by the cervical sympathetic and vagus. To give this method a fair trial it is best to place the anode by turns over the goitre, the heart, and over the abdomen. The current should be mild and continuous. Recently Rockwell has recommended the use of the faradic current.

As far as medicinal treatment is concerned the drugs influencing the heart's action have been most in favor; above all, digitalis and strophanthus. While these drugs act favorably in some cases they unquestionably exert an unfavorable influence in others. I have found that excessive palpitation of the heart could be controlled very much better by cold applications to the cardiac region, and by the use of the galvanic current, with the anode over the heart. I am willing, however, to concede that the effect of the latter may be due to suggestion. If there is great emotional excitement bromides in small doses may be given with advantage. Nitrite of amyl and nitro-glycerine have been recommended, but they are to be used with great caution, especially in children, and the result is a doubtful one at best. Ergot has been administered in some instances, and the subcutaneous injections of ergotin into the goitre have been practised, but the results have not been favorable enough to warrant its continuance.

Thyroidectomy, or excision of a part of the enlarged gland, has come into vogue, and has been reported by many German, English, and American writers. Improvement following the operation has been observed, and in my opinion is sufficiently marked to warrant further trial of this procedure; but I am not aware that the operation has, as yet, been attempted in children. Sufficient time has not yet elapsed to decide whether the relief obtained by the operation is merely temporary, or whether a permanent cure has been effected. If the enlarged gland presses upon

the trachea and menaces life, the operation is surely justified. Moebius, Leyden, and others have advised the use of Anti-thyroidin, Rodagen (prepared from the milk of goats whose thryoid glands had been removed); but the effect of these newer remedies is still sub judice—and great caution should be exercised in administering them.

THYROID ENLARGEMENTS AT THE AGE OF PUBERTY.—At this period the enlargement is often developed quite suddenly, and is so marked as to arouse suspicion of incipient exophthalmic goitre. As such girls are often anæmic, and have a high pulse-rate the suspicion of serious disease becomes still stronger; but such enlargement of the thyroid gland does not continue for any great length of time, and none of the other important symptoms of Graves's disease are developed.

TACHYCARDIA should also be mentioned, as it may occasionally give rise to the suspicion of Graves's disease. An excessively high pulse-rate is frequent in children, but true tachycardia in children has, to my knowledge, rarely been reported. I have seen several interesting cases of this kind, and one of them was of unusual interest. It was the case of a boy, of twelve years of age, who had been under my treatment for infantile cerebral hemiplegia, and had sufficiently recovered from this condition to be able to go about freely, to attend school, and to play with boys of his age. While playing on a cold winter's day he was struck in the neck by an icy snowball. Directly he felt faint, but did not lose consciousness. As soon as he was put to bed the tachycardia became evident, and his family physician was sent for. From the first day until about three weeks after the accident the pulse was at no time less than 200 per minute, and often exceeded this rate, so that the number could not well be counted. None of the drugs exhibited—such as bromides, strophanthus, digitalis, aconitia—had the slightest effect upon the heart; but after a period of about three weeks the symptoms subsided quite suddenly, and the boy, now a man, has been entirely well for many years. The injury to the cervical region, and possibly to the cervical sympathetic, causing tachycardia, is of interest, if we recall the part that the cervical sympathetic is supposed by many to play in the causation of Graves's disease. The treatment of such cases should consist of rest, and the application of an ice-bag to the heart and to the cervical spine; bromides, heart tonics, and aconitia may be tried according to the indications of the case.

MYXŒDEMA.

MYXŒDEMA is a form of trophic disease to which attention should be directed. The disease is of unusual interest, but we need not go into a full discussion of the subject, as the one form of it which occurs in children—myxœdematous idiocy—will be referred to in the Chapter on Idiocy. Myxœdema was first fully described by Gull, in 1873; four years later similar cases were reported by Ord, and since that time innumerable authors in

England, America, France, and Germany have studied this tropho-neurosis. Special interest has been attached to this disease, since Kocher described a condition following upon removal of the thyroid, resembling myxœdema, to which he gave the name, cachexia strumipriva. The Commission that reported to the Clinical Society of London, in 1888, agreed that myxœdema, cachexia strumipriva, sporadic cretinism, represented one and the same morbid entity. The condition following upon removal of the thyroid gland, the results of physiological experiments (implantation of the glands into the abdomen of animals — Schiff, Horsley, and others), and above all, the brilliant achievements of thyroid feeding, have proved conclusively that deficient action of the thyroid gland is the most important factor in the etiology of myxœdema.

FIG. 52.—Case of Myxœdema with Idiocy. Patient Twelve Years Old: Dwarfish in Stature.

Symptoms.—Of the symptoms of myxœdematous idiocy (the congenital form) we need here merely state that the skin is glossy or hard; the lips and tongue are thick and large; the hair is stiff and dry; the child is dwarfish in stature; the mental condition is that of marked idiocy or of imbecility. There is always absence of the thyroid gland.

Myxœdema resembling that of the adult may come on at the age of puberty, or even earlier. It is characterized by the following symptoms: A general increase in the bulk of the body; the skin is firm and inelastic, does not pit upon pressure, and is dry and rough; the folds of the skin disappear, and there is a general obliteration of all the lines of the skin, particularly in the face, giving, as a rule, an older and more stupid expression to the face; the nostrils and lips are very much enlarged. There is distinct apathy and slowness of speech, as well as of action, in some instances; the mental changes include delusions which lead to dementia. Albuminuria and glycosuria have been observed; but these are accidental complications, no doubt.

The disease cannot be mistaken for any other, except possibly chronic

nephritis; but the lack of pitting, the examination of the urine, and the peculiar expression of the face will help to remove all doubts.

The disease is slowly progressive, and may last for ten, fifteen, or more years. The prognosis was hopeless until the recent discovery of thyroid feeding, and success of this warrants us in rejecting for the present every other form of treatment.

Treatment is to consist of the administration of the pulverized thyroid gland of the sheep, which is to be had in all countries. Armour's preparation has, in my experience, been the most satisfactory. Patients taking thyroid gland should be observed carefully.

In children one grain twice a day should be the maximum dose at the beginning; this may be increased to three or five grains twice daily until the amount is established which the patient needs to keep in a normal condition.

A word of caution is in order not to continue the remedy if muscular pain and cardiac attacks occur. In several cases I have been compelled to discontinue the use of the gland on account of extreme and rapid emaciation. Tonic measures should be employed promptly if the general health of the child has been impaired.

ANGIO-NEUROTIC ŒDEMA.

ANGIO-NEUROTIC ŒDEMA is a trophic disorder, first described by Quincke, in 1882, although conditions closely resembling, or identical with it, have been known ever since 1827. It has passed under the designations of "acute circumscribed œdema," "periodic swelling," "Australian blight," "non-inflammatory œdema," etc.

The disease is characterized by the appearance of circumscribed swellings in various parts of the body, more especially in the face, throat, and the extremities. These swellings appear without any direct cause, and are often associated with gastro-intestinal disturbance, which is thought to be due to a condition of the mucous membrane of the stomach and bowels similar to that of the skin or larynx when these parts are diseased. The disease comes on in attacks, which are precipitated most easily by exposure to cold, and by slight or severe injuries. The attacks are most apt to occur between the hours of 1 and 5 A.M. In 71 cases the parts first affected were: The face in 29 cases; the extremities in 22 cases; the larynx in 5 cases; the genitals —penis, scrotum—in 3 cases; the trunk in 6 cases; the stomach in 3 cases; the gums, neck, ear, each in 1 case.

The œdema may be fully developed within one-half to two hours. The color of the skin is either whitish, or a dull roseate hue with a whitish shading near the centre of the œdematous patch. There is no pitting on pressure, and none of the signs of an inflammatory swelling. The swelling, if in the face, may be marked enough to cause disfigurement. The sudden jumping of the swelling from one part of the body to another is quite characteristic. Subjectively there is a sensation of scalding or burning, or an itching on the part affected; and after the œdema subsides a "heavy wooden" feeling is complained of. The disease is not a serious one unless it affects the mucous

membranes of the larynx and pharynx; if the œdematous swelling in the larynx increases rapidly, death may take place from asphyxia, as in two cases reported by Osler. One case has been reported that proved fatal from œdema of the glottis (Krieger).

The gastro-intestinal tract, if affected, may exhibit severe disturbance, such as pain in the epigastrium, vomiting, colic, retraction of the abdomen, and constipation or diarrhœa.

Œdema of the lungs has been reported as a symptom of this disease; but proof of this is wanting.

Collins has analyzed 75 cases.

According to this author the average age at the onset of the disease is twenty-seven; childhood is by no means exempt. Dinkelacker has reported the case of a child that had its first attack when three months old; similar cases have been reported by Widonitz and Goltz. The disease occurs nearly twice as often in males as in females. It may come on after any exhausting condition; but heredity, as well illustrated in a family described by Osler, is the most important factor. In this family five successive generations have been affected. Severe emotional excitement and hysterical conditions are the forerunners of the disease. The ingestion of certain foods was followed by this special form of œdema in cases reported by Osler, showing an etiological resemblance at least between angio-neurotic œdema and urticaria.

Matas and others have described cases of this disease coming on after or during malarial fever. The diagnosis is made readily enough; it may be confounded with the blue œdema of hysteria, as described by Sydenham, or the white œdematous swellings of hysteria described by Charcot; but the presence or absence of the stigmata of hysteria will help to disprove or corroborate the suspicion of angio-neurotic œdema.

There can be little doubt that angio-neurotic œdema is a vasomotor neurosis; it is analogous to the non-inflammatory swellings which Weir Mitchell described after injury to the peripheral nerves, and to similar swellings, which appear after stretching a nerve. I have seen œdematous swellings in the face, after section of the trigeminal nerve, very like the circumscribed œdema under discussion. The serous exudation in all probability results from a retardation of the blood-current, and this in turn must be attributed to a local paresis of the vaso-constrictors, or a reflex stimulation of the vaso-dilators.

Treatment of this disease as it occurs in children is more difficult than in the adult, in whom the removal of toxic causes, such as alcohol and tobacco, is all that is needed. In children the hereditary element is most marked, and all we can hope to do is to inhibit the attack and to strengthen the general nervous system. To check the attack we may compress the affected part by an elastic bandage, or apply dry heat. Small doses of morphia or codeia may be given. In the case of swelling in the pharynx or larynx, surgical interference may be necessary. To improve the general health of the child it will be best to administer strychnia in small doses (grain one-hundredth to grain one-sixtieth), and to give blood-tonics such as iron and arsenic.

RAYNAUD'S DISEASE.

RAYNAUD'S DISEASE, or symmetrical gangrene, is a very rare trophoneurosis, which occurs, however, quite as frequently in children as in adults. Indeed, says Morgan, "if there be any period when it is especially prone to occur, it is in childhood." Of 93 cases which he analyzed 24 were in children under ten years of age, and 5 of Raynaud's original cases were children between three and nine years old. Mendel observed a case in a child fifteen months old.

The symptoms of the disease are practically the same as those first enumerated by Raynaud in 1862. A localized ischæmia or asphyxia in symmetrically situated parts is the most constant symptom; this asphyxia may lead to gangrene, but does not invariably do so; nor is the disease invariably symmetrical, and for that reason the designation of symmetrical gangrene is not entirely appropriate. The order of development of the symptoms may vary somewhat. Before the local syncope is fully developed, there is often a general numbness and some pain in the part to be affected. The affected area becomes pale and waxy in appearance; if pricked, little or no blood flows from it. This condition may be recovered from, and several such attacks may occur, each one ending in recovery; finally, however, the condition persists, and local asphyxia is developed; in some instances the condition is reached at once and is persistent. The affected areas become deep red, then blue, and finally black in color; the parts are swollen, and the local temperature is lowered. Extravasation of blood into the surrounding tissues may occur. Recovery at this stage is still possible; but if the morbid process is continued for some time the parts grow thinner, the fingers become attenuated, and finally gangrene results. The gangrene comes on either some months after the first symptoms of local asphyxia, or it is developed at the earliest period of the disease. The tips of the fingers and toes may be destroyed by this process, or an entire hand or foot or some other considerable part may be destroyed by gangrene. The parts most frequently affected, are the fingers, toes, ears, the buttocks, the calves, and the nose. The gangrenous area is generally separated from healthy tissue by a sharp line of demarcation; the dead part may either become mummified, or it may be cast off after suppuration. Moist gangrene also occurs at times, with the formation of large bullæ and pus centres. If a gangrenous area heals, it behaves like a torpid ulcer (Elliot).

Intermittent hæmoglobinuria and glycosuria have been observed. The pathology of the disease is still obscure. Raynaud and his successors were of the opinion that the local syncope is produced by a contraction or spasm of the blood-vessels (both arteries and veins). The disease of the blood-vessels is by several authors attributed to syphilis or Bright's disease. The symptoms of Raynaud's disease have also been observed in association with tabes, syringomyelia, myelitis, and neuritis.

The prognosis is generally good; if death occurs, it is due to some intercurrent disease. Recoveries are frequent, even if local gangrene has

existed; but if large portions become gangrenous, the general health of the patient may be seriously impaired. In such cases tuberculosis may be developed and hasten death.

Treatment should be directed toward the improvement of the patient's general condition, and freedom from all emotional excitement should be insisted upon. The affected parts should be wrapped in cotton, and placed in a position most favorable for the circulation. Dry heat or warm baths may be employed to sustain the temperature of diseased areas. Nitro-glycerine and nitrite of amyl have been recommended, but they have not been exhibited successfully. Electric (galvanic) baths, and the application of the galvanic current through the cervical spine and the affected area have also been favored by some; but very little good has resulted from such treatment. The greatest benefit will be derived from surgical measures after gangrene has set in.

FACIAL HEMIATROPHY.

FACIAL HEMIATROPHY is a rare form of disease. Often it begins in childhood, and has important relations to other diseases of the nervous system. It has been described by some with exophthalmic goitre and with migraine, but its relations appear to me to be very much closer to scleroderma, which occasionally precedes it, and I have recently seen a case of universal scleroderma in which there was double hemiatrophy of the face.

The disease is characterized by a distinct diminution in the size and bulk of the subcutaneous tissues, and by the alteration in the bony structure as well. In one case, on the left side the distance from the middle of the chin to the angle of the jaw was 11 ctm.; on the right side, 12⅛ ctm.; from the upper margin of the naso-labial fold to the middle of the ear was 10¾ ctm., while on the right side this distance measured 11½ ctm. The first indications of atrophy are found in the skin. A single spot, generally in the cheek, becomes thin and white from the disappearance of pigment. The loss of fatty tissue underneath causes a depression also at this point. The atrophy proceeds from this and gradually involves the entire half of the face. The eye is retracted, there is distinct wasting of one-half of the nose, and even the ear of one side may be smaller than that of the other side. In some few instances a slight glossiness of the skin has been observed, but in the majority of cases there is unusual dryness, the condition resembling scleroderma. The hair also is apt to be stiff and hard. In some instances the teeth in the affected half are small, and undergo decay. There is no disturbance of sensation, and no interference with any of the special senses in the ordinary run of cases. The temperature sometimes varies a little. In a case which I reported the temperature was one degree higher in the normal ear than in the ear of the affected side. The muscular movements, in spite of the increasing atrophy, as a rule, remain normal. Several other authors, and myself, have reported cases in which there were distinct tonic and clonic contractions of the muscles supplied by the fifth nerve of the affected side. Atrophy of one-half of the tongue also occurs in some cases, but it is a rare complication.

The causes of the disease are unknown, except that several cases have been developed after severe injuries to the face. Thus in one case reported by Skryme, and quoted by Gowers, in a child three and a half years of age, the disease was developed six months after an accident in which "the face was jammed and the neck twisted." The disease has also been developed after acute infectious diseases, and after other causes which give rise to neuritis. There can be no doubt, however, that in many cases the disposition to the disease is congenital.

PATHOLOGY.—There was much dispute regarding the origin of this disease, some claiming that the cervical sympathetic was responsible, others making it a disease of the trigeminal nerve. There is no doubt that a condition very similar to hemiatrophy will follow upon lesions of the cervical sympathetic; but an autopsy performed by Mendel on a case which had been observed by many other men, proved beyond a doubt that in that case, at least, the disease was due to a proliferating interstitial neuritis of the left fifth nerve, and that this neuritis was most marked in the second branch; in that case the facial nerve was entirely normal, but the left musculo-spiral had undergone the same changes as the left trigeminal. My own case, in which there were marked clonic and tonic contractions of the masseters, would also tend to show that the trigeminus was the chief seat of the trouble, and that in some instances the motor, as well as the trophic fibres in the trigeminus can be involved in the same morbid process. The occurrence of a neuritis of other nerves, as well as the association of facial hemiatrophy with scleroderma, would seem to prove that the morbid process may in some cases extend beyond the fifth nerve.

The course of the disease is, as a rule, steadily progressive, though after the lapse of time there may be a complete standstill of all the symptoms. The disease does not call for any active treatment, excepting in cases like my own, in which the spasmodic contraction of the masseters was extremely painful and had to be relieved by opiates. The disfigurement of the face is the most serious feature of the disease. In the case of one patient I succeeded in rounding out the cheek again by inserting a light rubber plate, which was attached by a dentist to the patient's upper teeth.*

* In view of the excellent results obtained by thyroid feeding in a case of hemiatrophy with scleroderma, this treatment deserves a fair trial.

PART II.

ORGANIC DISEASES OF THE NERVOUS SYSTEM.

CHAPTER XII.

DISEASES OF THE PERIPHERAL NERVES.

Traumatism, toxic infections, exposure to wet and cold, rheumatism, the causes which lead to disease of the peripheral nerves in the adult are not frequent in children. A description will be given of those forms only which are commonly met with in the earlier years of life.

In diseases of the peripheral (mixed) nerves, whether there be an inflammatory or degenerative process, the symptoms are, as a rule, motor and sensory in character. The paralysis is limited to the muscles supplied by the diseased nerve and by branches coming off below the site of injury or disease. The sensory symptoms may be a hyperæsthesia or an anæsthesia, according to the degree of change. Paræsthesiæ are very common, and if the nerve is inflamed it is painful on pressure. It is a striking fact, however, that the motor fibres suffer much more readily than the sensory filaments; possibly because the former are farther removed from the nutrient cell, and probably because a collateral innervation between sensory fibres is more easily established. Sensory fibres are also regenerated more easily. Further symptoms are, marked atrophy, loss of reflexes in the diseased member, and changes in electrical reaction. The degree to which the last are developed gives an indication of the amount of degeneration in the nerve.

Trophic disturbances are very common in connection with peripheral neuritis and degeneration of peripheral nerves; but these disturbances do not, as a rule, appear except in severe cases. Local cyanosis, œdema, and ulcers are common; the skin becomes smooth and glossy, and the growth of the nails may be interfered with.

In children, as in adults, the nerves of the brachial plexus are most frequently affected.

The symptoms referable to lesions of the brachial plexus vary according to the part of the plexus involved. As in the adult, we can distinguish between several distinct types; the upper-arm type, the lower-arm type, and the paralysis due to total plexus lesion.

THE OBSTETRICAL PALSIES.

As the name implies, these palsies are due to manipulations during labor. If the shoulder and clavicle are pressed

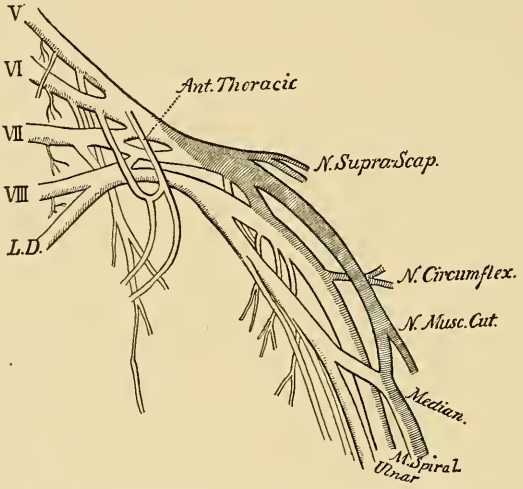


FIG. 53.—The Brachial Plexus: the Branches involved in the Upper Arm Type of Paralysis are indicated by Shading.

backward and upward injury to the plexus may result therefrom. In cases of breech presentation pressure upon the supra-clavicular spaces may cause a partial plexus paralysis. I have seen several cases in which this paralysis was bilateral. The disturbances that ensue upon these obstetrical manipulations may be the result of a simple traction upon or of laceration (Pierce Clark) of some or of all the fibres of the brachial plexus (Erb's type most frequent).

The affected arm hangs down limp. Although the child moves the fingers and the arm of the normal side, there is little or no power on the paralyzed side. In the first

few weeks after birth, if electrical examination is attempted, it will be found that the muscles of the affected extremity do not respond to the electric currents. At later periods, in the milder cases, the galvanic response of the nerves may be a little altered, but the muscles, as a rule, do not exhibit any marked change. If the case is a more severe one the diseased branches of the brachial plexus will fail to respond both to the galvanic and faradic current, and the muscles supplied by these branches will show either an altered galvanic response, the anodal closure contractions being greater than the cathodal closure contractions, or the affected muscles may fail to respond altogether to the intensity of current which can be employed in children. The ordinary tests for sensory disturbances cannot easily be applied in very young children. There is generally some slight diminution in all forms of sensation, but we can only state with certainty that the sensations of touch and of pain are impaired in all but the mildest cases. In considering the imperfect sensation of a very young child we must bear in mind that, as Soltman and Westphal have shown, the peripheral nerves are not fully developed until several weeks after birth. Evidently the entire central and peripheral apparatus conducting sensation is developed later than that transmitting motion.

The appearance of the affected limb changes a little as time goes on. In some of the milder forms of obstetrical paralysis a complete restoration takes place within a few weeks, or a few months, with or without medical interference. In the severer forms the atrophy is apt to increase; the development of the limb is retarded as compared with the normal side. It is common enough to have contractures at the elbows and at the wrists develop in the course of a few months, or after a year or more, in those cases in which, either from lack of treatment or from the severity of the injury at birth, the initial paralysis remains unaltered.

Pathology.—It is naturally a difficult matter to determine the exact changes which take place in any part of the brachial plexus in consequence of obstetrical manipulations, for the opportunities of a post-mortem examination are very rare. Roger describes a case in which the facial

nerve, and the arm of the same side were paralyzed immediately after birth from pressure of the forceps. Large extravasations of blood were found post mortem in the vicinity of the stylomastoid foramen, and of the brachial plexus. Oppenheim, who examined one of Henoch's cases, discovered degenerative changes in the brachial plexus. In the milder forms of these peripheral lesions we must suppose that the disturbance of function was due to slight mechanical injuries, to a stretching or tearing of some of the fibres, possibly to slight injury of the nerve-sheath, or a mild form of inflammatory reaction in the nerve-tissue. In the severer cases the lesion may amount to an actual tear, to a severance of the nerve-fibres, or, in the case of subluxation of the head of the humerus, to actual compression, with loss of function, of the nerve-fibres. The lower arm type (Klumpke) of brachial plexus lesion has been reported. Also a case of laceration of the spinal roots (Philippe and Cestan).

DIAGNOSIS.—The differential diagnosis is to be made between these obstetrical palsies and the so-called "birth palsies" due to cerebral lesions. A knowledge of the symptoms due to lesions of the motor tract will be useful.

PERIPHERAL PALSIES (OBSTETRICAL PALSIES OF BRACHIAL PLEXUS).	CEREBRAL BIRTH PALSIES.
1. Arm only affected.	1. Hemiplegia or diplegia common; brachial monoplegia rare.
2. Flaccid paralysis with atrophy.	2. Spastic paralysis, with or without atrophy, with tendency to rigidity.
3. Deep reflexes absent, surely not exaggerated.	3. Deep reflexes increased.
4. Changes in electrical reaction from simple loss of faradic response to complete reaction of degeneration.	4. No changes in electrical reaction.
5. No convulsions.	5. Convulsions apt to occur and to be repeated.
6. Deformity and arrested growth of entire extremity.	6. Flexion contraction of fingers, wrist, and elbow.
7. Sensation may be impaired.	7. Sensation not affected.

There is no other condition from which these obstetrical palsies need to be differentiated if the child is examined within a reasonable time after birth; but if, as is so often the case, the child is brought to a physician when it has reached the age of six, seven, or even twelve years of age, the true cause of the palsy may be difficult to fathom. At my clinics I have often seen cases for which nothing had been done, and the cause is elicited only after the closest inquiry from the mother. In older children it is a question whether the palsy dates from birth, or whether some accident in the first few years of life may not have been the cause. The pulling of the arm in some of the games the children play, wrenching and twisting it as they do with considerable force, may result in palsies very much like obstetrical palsies. Poliomyelitis is not easily to be confounded with these obstetrical palsies, for the former is rare in the first year of life, and relatively rare in the upper extremities; the atrophy is generally more extreme, sensation is not involved, and complete recovery is not common in the spinal paralysis of children.

Prognosis.—If properly cared for, recovery may be expected in all but the most severe forms of obstetrical peripheral palsy. According to the severity of the symptoms, the length of time that it will take before recovery sets in can also be foretold, with some degree of certainty, and for this purpose the electrical behavior of the nerves and muscles constitutes a fair test for the amount of damage that has been done. Even though the paralysis be complete, if the faradic response of the branches supplying the affected muscles is not entirely lost, the probability is that complete recovery will set in within a period of two to three months. If the faradic response is lost, but the galvanic formula is not altered, the restoration of function may be expected within a period of six months; but if both the faradic and the galvanic responses are entirely gone or seriously impaired, it may be a year, or even two years, before the arm can be properly used. In the severer cases, in which a great deal of injury has been done to the brachial plexus, it will not be well to promise much regarding the time or the possibility of complete recovery. Some of these cases

are extremely stubborn; this is to be emphasized, for the majority of authors seem to regard these conditions too favorably. Much will depend upon the accuracy of treatment. One point which I wish to impress upon the reader, however, is that in a fair proportion of all these cases the physician may reassure the parents regarding the future condition of the paralyzed extremity.

TREATMENT.—If the child is seen immediately after birth, and the diagnosis has been properly made, the best the physician can do is to let the limb severely alone for a period of at least two weeks; at the end of that time the exact damage that has been done can be properly estimated and the necessary therapeutic measures can be employed. Wrapping up the arm carefully, so as to avoid any further injury to it, and allowing it to be bathed as the rest of the body is, will be all that is necessary. After a period of two weeks I would advise the use of light friction and massage, so as to keep up the nutrition of the muscles, and I am also in favor of directing an intelligent nurse or relative to move the parts systematically so as to overcome any tendency to contracture. After a period of four to six weeks electricity may be employed, simply as a mode of exercise. If the parts respond to a mild faradic current such may be employed, but if the faradic current fails to elicit a response it is sheer waste of time to employ this form of electricity, and it will be better to use a mild galvanic current, using that pole which will produce contractions. Sittings of from five to ten minutes every day, or every other day, are quite sufficient, and will be of some benefit in the way of preventing excessive atrophy and of preventing the formation of contractures.

As soon as there is a tendency to the development of contractures, it will be well to place the arm in a splint in such a way as to oppose the contracture by simple mechanical force, and to take the arm out of the splint daily for the purpose of treating it by proper massage and electricity. If deformities have arisen as the natural result of disease, or from neglect, the child should be placed in the hands of an orthopedic surgeon and an attempt should be made by every known surgical procedure to correct the

existing trouble. The old belief that a child will outgrow such deformities is absolutely unfounded. It is an unfortunate fact, but true, nevertheless, that children outgrow only the very mildest disturbances of the nervous system, which is equivalent to saying that some troubles undergo spontaneous cure. Clark and others have advised suturing of the lacerated strands in severe cases; some favorable results have been reported. Great caution should be exercised in the selection of cases.

Paralysis of the lower limbs due to obstetrical manipulations is reported every now and then, but it is extremely rare. The lumbar and sacral plexuses are so well protected at every point of emergence from the spine and in their course through the gluteal and pelvic regions that they are not easily disturbed by any amount of manipulation or traction upon the legs. An injury of these parts is, however, conceivable. If present, the paralysis will resemble the paralysis of the adult in the number of muscles involved and in the character of the palsy. It is interesting in this connection to note that Ross, many years ago, supposed that traction of the legs was occasionally responsible for spastic paraplegia in children. He does not refer to the effect upon the nerve-roots, but supposes that such traction would be followed by derangement of the fibres in the spinal cord itself. This view has not been accepted, and it has been shown that cases of spastic paraplegia due to traumatism during birth are due, in the vast majority of cases, to cerebral and not to spinal injuries. Some exceptions have been credibly reported.

Still another obstetrical palsy occurring quite exceptionally is facial paralysis due to compression of the peripheral portion of the nerve by the blade of the forceps. This is an unusual occurrence, for if the blades of the forceps are properly applied they should not be in the vicinity of the facial nerve; but accidents will happen and the blades occasionally slip even if applied by skilful obstetricians, and thus cases of obstetrical facial nerve palsy are not entirely unknown. This may be the result of actual pressure, and in other cases an extravasation of blood into the region of the parotid is likely to compress the nerve sufficiently to cause temporary palsy. Exostoses within the pelvis or hæmatomata may cause facial palsies by direct pressure (Vogel, E. Kehrer, Schultze). Schütze and A. Stein have reported facial and hypoglossal palsy, due, presumably, to the rare coincidence of an external hæmatoma and a small hemorrhage into the hypoglossal nucleus (during normal labor).

The prognosis in these cases is, on the whole, favorable. In a few cases, however, a degeneration of the nerve occurs in consequence of the severity of the compression. Henoch reports two such cases. Parrot and Troisier have examined such nerves post mortem. I have the records of two cases, one in a girl of twelve, the other in a woman of some twenty-two years of age, in whom facial palsy with unequal development of the two halves of the face is the result of this obstetrical accident.

The treatment of these cases is exactly the same as that for all other peripheral palsies. Light massage and the use of the electric current are the only measures that can be safely recommended.

FACIAL PALSY (BELL'S PARALYSIS).

Disease of the seventh nerve is not infrequent in children from the same causes that result in facial paralysis in the adult. We have to consider, first, rheumatic facial palsy, more properly perhaps refrigeration palsy; second, facial palsy due to ear disease; and third, facial palsy as an accompaniment of disease at the base of the brain.

Rheumatic facial palsy may occur in children at almost any age. Although I have seen it in a child nine months old, it is rare before the age of three years, and is most frequent between the ages of six and fifteen years. Sudden draughts of cold air are by far the most frequent cause. The side of the face exposed to the draught is generally the one that is paralyzed. In a few instances it is difficult to make out the exact cause, but even under such circumstances refrigeration is very probable. I am not aware that facial palsy occurs in connection with the various diatheses in children as it does in the adult, in whom it is met with in association with diabetes or tuberculosis.

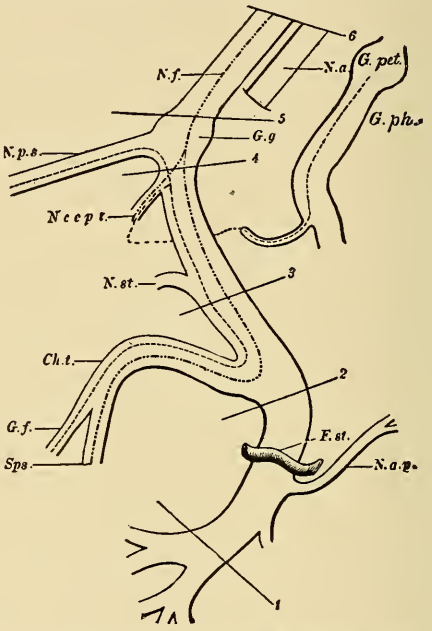


FIG. 54.—Diagrammatic figure showing the Relations of the Seventh and Glosso-Pharyngeal Nerves and the Course of the Taste Fibres. The lines 1, 2, 3, 4 mark off different segments of the nerve. *N. f.*, facial nerve; *N. a.*, Acoustic nerve; *G. ph.*, glosso-pharyngeal nerve; *G. g.*, geniculate ganglion; *N. p. s.*, great superficial petrosal from fifth nerve; *N. c. c. p. t.*, tympanic plexus; the dotted line *S. p. s.* indicates salivary fibres; the other dotted line indicates taste fibres; *Ch. t.*, chorda tympani; *N. st.*, stapedius nerve; *F. st.*, stylo-mastoid foramen. (Modified from Erb by Dana.)

The symptoms of facial palsy will be readily understood by an examination of Fig. 54. The distal portion (1) is the one which is generally affected in these rheumatic cases. All the branches of the facial nerve are involved, but there is no interference with taste, with hearing, with movement of the palate, or with the secretion of saliva. The paralysis involves the muscles of the eyes, the nose, the cheeks, and the lips. On inspection of the child's face it is easy to make out which side is affected, as the eyelids of the diseased side are wide open (lagophthalmos). If the child is asked to close the eyes it does so with ease on the sound side, but the diseased side fails to respond.* In this effort there is over-action of the frontal muscles and of the corrugator supercilii on the sound side; the diseased side showing either diminished action or a total paralysis. The child cannot pull up its nose; in blowing out the cheeks the diseased side is flabbier than the sound side, and if the attempt is made to whistle or to pout the lips, as in kissing, the insufficient action of the muscles on the diseased side becomes very evident. If asked to show the teeth, the muscles of the sound side overact, and the mouth is drawn toward the healthy side. By this movement the paralysis is often revealed, even if the parts appear perfectly normal while at rest. The tongue when protruded deviates toward the sound side; this deviation was formerly considered to be apparent, not real; but Hitzig has shown that there is an actual deviation toward the sound side, and supposes that this is due to the fact that the tongue wishes to avoid contact with the corner of the mouth on the paralyzed side. Contractures of the muscles of the paralyzed side occur in the later stages of a facial palsy. The lower branches recover, as a rule, more slowly than the upper, and in several cases I have observed that the lower and middle muscles could be contracted only if the attempt was made to close the eyes (Fig. 55).

Sensory disturbances are entirely absent, as the facial nerve is a purely motor nerve, the sensory fibres of the face

* A few cases of double facial palsy have been reported (Mott, Huebschman); a successive involvement of the two sides is more frequent. The author has observed it in cases associated with diabetes in the adult.

coming from the fifth nerve. I have seen a few cases of severe neuralgic pains complicating facial palsy in children, but to account for it there has always been some additional cause, as in a boy of fourteen, whom I recently examined, in whom decayed upper teeth offered the explanation for the neuralgic supraorbital pain.

Whatever part of the nerve is involved, provided it be between the nucleus of the facial nerve in the pons and the periphery, electrical changes will be found. In all but

FIG. 55.—Boy with Facial Palsy, in Stage of Recovery. Slight contracture of paralyzed side (B); in figure A mouth is pulled to the paralyzed side when boy attempts forcibly to close left eye; also deepening of naso-labial fold.

the very mildest cases the faradic response of the nerve as well as of the muscles is lost. My own experience has proved, however, that there is an exception to this rule, and that is, that if the patient be examined within the first forty-eight or seventy-two hours after the onset of the palsy, the faradic response may still be present; it rapidly diminishes, however, and after a period of three days, in the vast majority of cases, it is entirely lost. The galvanic response of the nerve may be increased during the first few days, but it is soon diminished or lost. The galvanic irritability of the muscles supplied by the facial nerve shows many interesting changes. In the earlier stages of the disease, and

in all but the severest cases, there is an increased galvanic excitability for a period of about two weeks. The same currents which produce very strong contractions on the diseased side give weaker contractions on the sound side. This can be easily demonstrated in the case of the orbicularis oris. If a very weak current be applied to the sound half of the muscle, contractions may be seen in the diseased half and not in the normal half, the increased excitability in the latter being so great that the small amount of current diffusing to the paralyzed side is sufficient to produce a contraction, while the full current is not sufficient to make the healthy half contract. During this period the cathodal closure contraction of the diseased muscle is generally greater than its anodal closure contraction. After a little while the increased excitability diminishes and soon falls below that of normal muscular tissue. The polar contractions also indicate a change, the cathodal closure contraction first being equal to the anodal closure contraction; later on in the severer cases the order is entirely reversed.

As the disease approaches the period of recovery the galvanic formula approaches more nearly to the normal, the cathodal closure contractions being equal to the anodal closure contractions. Finally, the cathodal closure contraction becomes greater than the anodal contraction, and the contractions that have been sluggish once more become prompt and short. The faradic response is, as a rule, rather longer than this in returning, and it is not uncommon to see the full power of all the muscles return before the normal electrical reactions have been established. In the case of a boy (Fig. 55) the faradic response of the muscles and a normal galvanic formula were not restored until several months after the contractures had set in, and after most of the muscles could be innervated tolerably well.

In facial palsy due to ear disease, the symptoms can be easily accounted for by the position of the nerve in the Fallopian canal. The nerve may be involved during an attack of otitis media, and as this ear affection occurs often enough in children in association with several acute infectious diseases, notably scarlatina, this form of facial palsy is quite a frequent occurrence. It does not

occur as readily in the earlier stages of an otitis as in the later suppurative stages, when, in consequence of caries of the bone, the facial nerve is directly involved. According to the severity of the disease the paralysis will be more or less complete, and as far as the facial nerve is concerned the symptoms will resemble those of the rheumatic form, but naturally the association of facial palsy with defective hearing or the persistence of a purulent discharge from the ear will point to the actual cause of the paralysis.

I have seen facial palsy occur also after mastoid operations for ear disease. In these cases an accident during the operation is often sufficient to break through the fine bony plate separating the facial nerve from the ear structures, and a more or less severe facial palsy is the result. This occurred in a little patient whose case I reported fifteen years ago, in whom the mastoid operation was done for the relief of epileptic attacks which were caused by the retention of pus. On his recovery from the operation a distinct facial palsy on the side of the operation was noticed, and existed for a period of very nearly three months, after which complete recovery took place.

As in the adult so in the child, paralysis of the face may accompany lesions at the base of the brain and in the pons. If the lesion is at the base, other cranial nerves are involved as well as the facial, and the diagnosis can therefore be readily made. It is common enough to find the facial nerve partially or totally paralyzed in cases of basilar meningitis, whether tubercular or non-tubercular. In cases of injuries to the skull, or of tumors occupying the middle or posterior fossa, facial palsy is one of a large series of symptoms and has but little diagnostic value excepting in its association with affections of other cranial nerves. It may be stated in this connection that in cases of cerebellar tumor, which are by no means rare in children, both the acoustic and the facial nerves, which lie in very close juxtaposition to each other and are easily pressed upon by a cerebellar growth, are often paralyzed and give rise to symptoms which are almost pathognomonic of cerebellar disease. If the lesion is in the pons, and particularly if it be below the decussation of the facial nerves, paralysis of the face is

associated with alternate hemiplegia. In these cases every branch of the facial nerve is apt to be involved. If the lesion be a neoplasm other symptoms pointing to compression of the cranial nerves and indicating the presence of tumor within the cranial cavity will naturally be associated with those mentioned before.

The diagnosis of facial palsy is easy to make, and the only difficulty that arises at times is to determine whether the palsy is indicative of a central or a peripheral lesion; but this difficulty can be readily met by keeping in mind the fact that if the lesion is in the brain anywhere above the pons the upper branches of the facial escape altogether and the electrical responses remain entirely normal; furthermore in such a case the facial palsy is associated with hemiplegia on the same side of the body as is the paralysis of the face. It is far more difficult to determine the exact part of the nerve that is diseased. As the intensity of the affection depends somewhat upon the extent of the nerve tract that is diseased, this inquiry into the accurate localization of the disease has a practical as well as a scientific interest. Taking the diagram on page 216 we can study the variation of symptoms according to the part that is affected. If the disease is limited to that part outside of the Fallopian canal, there is complete paralysis of every branch of the facial nerve with the exception of the posterior auricular. Reflexes are wanting, but there are no disturbances of taste or hearing, no paralysis of the palate, and no disturbances of salivary secretion. If the disease happens to affect the part of the facial nerve as far as, but not including, the chorda tympani (Fig. 54, line 2), the symptoms are paralysis of all the branches of the facial, including the posterior auricular.

The same symptoms as the above, with the addition of a disturbance of taste in the anterior portion of the tongue and a diminution in salivary secretion points to an involvement of the facial nerve from the point of junction of the chorda tympani to the junction of the stapedius with the main trunk (from 2 to 3).

If that part of the nerve lying between the stapedius and the ganglion geniculi is diseased, the symptoms, in addition to total paralysis, are changes in taste, diminution in salivary secretion, and abnormal acuity of hearing, but there is no paralysis of the soft palate. The above symptoms, with the addition of paralysis of the soft palate, point to a lesion in the region of the geniculate ganglion (division 4 to 5); but on this point there is much doubt. See table, p. 13.

Paralysis of all the peripheral branches, as well as paralysis of the soft palate, abnormal acuity of hearing, decreased salivary secretion, but no involvement of taste, help to locate the lesion above the geniculate ganglion, and between it and the exit of the facial nerve from the brain (division 5 to 6). If this part of the facial tract is diseased other cranial nerves are frequently affected. In such cases we are apt to have paralysis of the abducens, loss of hearing on the same side as the facial palsy, tinnitus aurium, and the reaction of degen-

eration of the acoustic nerve. If we have total palsy of every branch, palsy of the soft palate without disturbance of taste and with simple diminution of electrical excitability, without any other marked symptoms, we may locate the lesion in the facial nucleus; but this form of palsy is recognized more definitely if other cranial nerves are simultaneously affected, such as the hypoglossal, the spinal accessory, the vagus, the trigeminal nerve, etc.

The association of the symptoms just mentioned with paralysis of the arm and leg of the opposite side, puts the lesion in the pons. If there is but partial paralysis of the face, if the electrical excitability is not affected, if the reflexes are normal, and if the extremities of the same side are affected, the lesion is in the crus or in the hemispheres, say in the internal capsule. If the facial nerve and the oculo-motor nerve of opposite sides happen to be affected the lesion is positively in the crus on the same side as the ocular palsy.

The course and prognosis of all forms of facial palsies will depend entirely upon the nature of the morbid process. If the disease is due to caries of the petrous portion of the temporal bone, to malignant tumor, to fracture, to tubercular meningitis, or to any form of meningitis, the prognosis is extremely grave. Even those cases depending upon specific disease at the base are not as amenable to treatment as one might expect. Those due to parotitis or to internal otitis recover if the disease itself is not of the destructive type. Cases due to simple section of the nerve, as in cases of operation, yield a tolerably favorable result.

The prognosis of the rheumatic forms of facial palsy varies according to the intensity of the disease, but it does not depend apparently upon the amount of nerve-tissue that is affected, for, on the whole, cases in which the nerve within the canal is involved take about as favorable a course as do those in which the pes anserinus is the only part affected. Erb distinguishes three forms—a mild form, a moderate form, and a severe form of facial paralysis. His division depends entirely upon the electrical conditions present at the end of the first week. I cannot agree altogether with his statement that if the electrical excitability is entirely normal at the end of the first week, as in light forms, recovery will set in in two or three weeks. If at this same period the faradic and galvanic excitability are slightly diminished recovery may be expected in from four to six weeks, and if at the end of the first week the elec-

trical excitability of the nerve is very much diminished or totally lost, the disease will run a course of many months. I have seen cases get well practically within four weeks, in which the faradic and galvanic excitability of the nerve was diminished very much from the very start, and I should be inclined to formulate the following statements:

I. If at the end of the first week, or, still better, at the end of the second week, the nerve responds at all to the faradic or galvanic current, a prompt recovery in about four weeks may be expected.

II. If at about the same time the nerve fails to respond, but the muscles show a diminished or altered galvanic response, the disease is likely to run a course anywhere between one and three months.

III. If the muscles respond but feebly to strong currents, if the galvanic formula is altered, and if the contractions are extremely slow, the disease may run a course anywhere between six months and a year, or even longer. If after a period of two months no electrical reaction can be observed, the degeneration is very complete, and a paralysis lasting at least a year, if not longer, may safely be predicted.

Treatment.—We need say little of the treatment in cases in which the facial palsy is only one of many symptoms pointing to gross cerebral disease. Under such circumstances treatment directed to the relief, or possibly the cure, of these conditions will have to be employed. In the cases of facial palsy due to ear disease, proper surgical treatment should be recommended at a very early day. Much doubt has been expressed as to whether any treatment can curtail or cure the rheumatic palsies. The milder forms will undoubtedly get well even if no treatment is attempted. In the more chronic cases the use of the electrical current, particularly of the galvanic, can be safely recommended, for I have the distinct impression that cases so treated, and particularly those that have been properly treated from the very start, run a somewhat shorter course than those in which no electrical treatment has been attempted.

In the year 1893 Remak gave statistical evidence in favor of this view, and put a quietus for the present upon those

who think that there is nothing in electro-therapeutics save the effect of suggestion.

The electrical current, even if it have but slight influence in restoring the power of conduction in the diseased nerve, is surely of value, as it is in paralysis of the extremities, in exercising parts that cannot be moved by the will, and for this reason if for no other it deserves to be employed.

Counter-irritation has been repeatedly tried. This may be accomplished by blistering either with emplastrum cantharidum just in front of the ear or over the mastoid, or by the use of the actual cautery. I am free to confess, however, that the value of counter-irritation seems questionable in all cases, and particularly in those in which counter-irritation is not indicated to allay pain that is so frequently associated with neuritis. The application of leeches is to be thoroughly condemned, except possibly in those cases in which the facial palsy is associated with ear disease. This practically limits us to the use of electricity as the only therapeutic measure which promises some relief in the severer cases. In these cases the faradic current, as recommended by Erb and Duchenne, may be employed for the first week or two, but after that a stabile galvanic current applied over the mastoid and along the peripheral divisions of the facial nerve is more in order. As soon as the excitability of the nerves and muscle is increased, or shows an approach to the normal, regular electrizations of the paralyzed muscles, in sittings of ten minutes or more, two or three times a week, should be practised. I wish to caution the physician, however, against the use of strong currents or against the use of the electrical current without the safeguard of a galvanometer. A current varying between two and four milliampères is quite sufficient for all purposes. I have also directed my patients, as soon as the least power returns, to practise contraction of the muscles (closing of the eyes, pouting of the lips, etc.), while looking into a mirror.

As for medicinal measures, they may be discarded altogether. In many cases I have tried strychnia both per os and in the form of hypodermic injections, and I am firmly convinced that the effect of such medication is absolutely

nil, nor can we expect much more from the use of irritating ointments or douches to the face. If drugs must be administered one may exhibit iron, arsenic, and cod-liver oil. In inveterate cases, union of the facial with the spinal accessory or the hypoglossal nerve may be attempted.

Other Peripheral Palsies.—Almost any peripheral palsy which is known to occur in the adult may at times occur in the child. Of these I will make special mention only of a peripheral palsy, generally of traumatic origin, affecting the peroneal nerve. Injury to this nerve occurs readily enough if a child happens to be violently struck upon the leg, or if the leg happens to be squeezed against a post, a chair, or, as in one case I have seen, against the edge of the bed. I mention this special palsy because it may lead to slight difficulties in diagnosis, inasmuch as confusion may arise with acute anterior poliomyelitis which affects the muscles supplied by the peroneal more often than those supplied by any other nerve.

The onset of the paralysis without fever, the presence of considerable pain, the marked and rapid tendency to recovery, and the history of violent traumatism will help to distinguish this peripheral palsy from the essential paralysis of children. The course, prognosis, and treatment of such a peroneal neuritis will be exactly the same as in the case of other nerve palsies.

Spasm of various peripheral nerves is common in young persons. The facial and the spinal accessory nerves are frequently affected; the trigeminal in rare instances only. Spasm of the facial nerve (tic convulsif) is much less frequent than in adults; a spasm involving an entire facial nerve of a child is very rare indeed; but partial spasms are observed in the form of nictitation (clonic spasm) or of blepharospasm (tonic spasm of the obicularis palpebrarum). These movements are generally due to the influence of cold, to irritation of the trigeminus, but are also frequently of psychic origin. In the cases of clonic spasm of the entire nerve, all sorts of extravagant grimaces are indulged in. These spasms must not be confounded with habit chorea.

Spasm in the distribution of the spinal accessory nerve involves either the sternocleido-mastoid or the trapezius (spasmodic wryneck).

The spasm may be reflex in origin, or due to irritation of the nerve in its course from the medulla and spinal cord to the periphery. The position and movements of the head will vary according to the muscles involved. These spasms may be unilateral or bilateral.

If we remember that spasm, whether clonic or tonic, denotes an excess of normal function, the symptoms can be easily understood by reference to the tables on pages 15 and 16.

Ordinary wryneck, so frequent in children, is due to a rheumatic myositis. Symptomatic wryneck occurs in connection with cervical adenitis, abscesses and caries of the cervical spine. Congenital wryneck is due to some pre-natal disturbance; it may be of intra-uterine origin, and denote imperfect development, in which case it is generally associated with atrophy

of one-half of the face; at times it is due to obstetrical injuries in cases of breech and foot presentations.

The treatment of spasms of the peripheral nerves consists in the use of sedatives, such as opium, conium, codein, chloral, and bromides; in some cases the valerianate of zinc, or the ammoniated tincture of valerian, might be given. The galvanic current may be applied in the milder cases. The attempt may be made to check the spasms by hypodermic injections of hydrobromate of hyoscine, one two-hundredth to one one-hundredth grain, or by the use of atropia (one-fiftieth to one-tenth grain, as recommended by Leszynsky).

Tenotomies are in order in the congenital and chronic cases. Keen has suggested resection of the posterior branches of the upper three or four cervical roots; but the advisability of this procedure is still in doubt.

Spasm of the hypoglossal nerve I have not seen in children. The complex co-ordinated spasms have been considered in Chapter VI.

CHAPTER XIII.

MULTIPLE NEURITIS.

NEUROLOGY has few more signal achievements to show than the advances that have been made in the study of multiple neuritis within the past decade. Many conditions which were formerly supposed to be due to an affection of the spinal cord, and which were often confounded with poliomyelitis, and with various forms of acute and subacute myelitis, are now positively known to be due to polyneuritis. The term multiple neuritis or polyneuritis is used to embrace all those diseases in which the symptoms are due to disease of several or many of the nerves of the body. There is not only a tendency to multiplicity but also a tendency to symmetrical development.

Many of the older writers recognized the possibility of multiple nerve affections, and a multiple neuritis, as such, was recognized by Dusmenil as far back as 1864, and even a few years earlier Virchow had described a multiple neuritis occurring together with leprosy. Going back still further, the historical investigations have led to the discovery that Dr. Jackson, of Boston, had recognized peripheral multiple neuritis as early as 1822; but all these researches, even including those of Duchenne, did not help to place the knowledge of multiple neuritis upon a firm basis. The subject was revived by Joffroy, in 1879, and Leyden, in 1880, and since that time innumerable articles have appeared by writers of almost every nationality.

Comparatively few writers, however, have had much to say regarding multiple neuritis occurring in children. Diphtheritic paralysis, to be sure, was discussed clinically long before it was known that it was due to peripheral neuritis. Multiple neuritis is not so frequent in children as in adults, for the simple reason that the causes leading to it are not as powerful in early life as later on; and yet cases are common enough to necessitate a discussion of them in this book.

SYMPTOMS.—Whatever the cause of the multiple neuritis may be the symptoms are, with few modifications, practi-

cally the same in all cases. Though the development of the various symptoms is subject to great variations, the symptoms are such as we might well expect on physiological grounds from disease of the peripheral mixed nerves. The

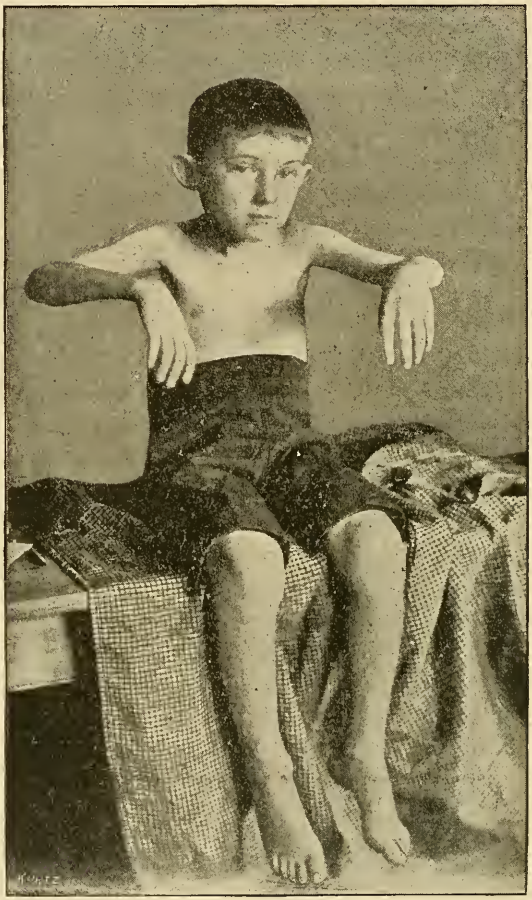


FIG. 56.—Young Boy with Multiple Neuritis, showing Double "Wrist Drop" and Slight "Foot Drop."

most characteristic feature of multiple neuritis is the association of motor paralysis with sensory paralysis, the distribution of each harmonizing with the other and showing very definite anatomical limits. The paralysis is of the flaccid order, leads at an early date to atrophy of the af-

fected muscles, and the electrical conditions are altered so that we may find almost every possible form of the reaction of degeneration from a mere loss of faradic irritability to an absolute loss of galvanic response on the part of the nerves and muscles. The distribution of the paralysis is, as a rule, entirely symmetrical, and may affect either the upper or lower, or all four extremities; it may involve every part of all the extremities, and is the one affection which, perhaps more frequently than any other, leads to a complete paralysis of every limb of the body. It is one of the diseases, therefore, which the physician should bear in mind if he finds a patient who is totally paralyzed with the bare exception of the head and tongue, the jaws and eyes; sometimes some of the latter parts may be paralyzed as well. Certain groups of muscles are more readily affected than others; thus we find that the extensors of the wrist and the extensors of the feet are very frequently involved, and at an early period of the disease "wrist-drop" and "foot-drop" are characteristic phenomena of the disease. With the paralysis sensory symptoms go hand in hand; paralysis associated with pain may be noted in some cases of spinal-cord disease, as in the acute stages of a poliomyelitis, but the persistence of pain in those parts which are paralyzed, and along the nerve-tracts, points very strongly to a multiple neuritis. Whenever we suspect that a paralysis or a deformity may be the result of a multiple neuritis, the previous occurrence of a painful affection and the duration of the same must be determined with the greatest care. If the patient is seen during the acute stage of the disease the tracts of the peripheral nerves will be found painful, and this objective proof will be added to the subjective symptoms which the patient describes. But the absence of pain, as evidenced by cases reported by Barrs, need not militate against the diagnosis.

The motor and sensory paralysis is not developed as rapidly as is the case in central affections. There is a premonitory period during which numbness, tingling, sensations of heat or cold, in the parts to be affected later on, are the annoying symptoms, and after a premonitory period that may vary from a few days to a few months the development of the paralysis may be acute, subacute, or chronic. In the majority of the cases it is sub-

acute or chronic, and it often requires a period varying between one and two weeks until the weakness of the parts has advanced far enough to be considered an actual paralysis. The onset is marked occasionally by the occurrence of fever, and if the disease is due to any toxic agent that has constitutional effects the beginning may be signalized by convulsions as well as by rigors. Under the latter circumstances the symptoms may very closely resemble those of an acute poliomyelitis, but it is rare, after all, to find as stormy an onset in cases of multiple neuritis as in spinal infantile paralysis.

Before the paralysis becomes absolute, if it ever reaches that stage, tremor of the parts and inco-ordination may be associated with loss of power and loss of sensation. The patient may be able to stand or walk, but does so imperfectly, sways with the body when attempting to stand, and may fall if the eyes are closed. The reflexes connected with the parts affected by the paralysis are almost invariably diminished or lost; this is particularly true of the knee-jerk, since the trouble so often begins in and involves the legs to a greater extent than any other part of the body. The reflex is often absent before the paralysis is developed. An exception to the rule of loss of knee-jerk has been reported by Strümpell. A few years ago he described a case in which the reflexes were increased, and this increase he attributed to irritation of the afferent portion of the reflex arc.

The more distal parts of the extremities are, as a rule, the first to be affected. In the lower extremities the earlier stages of the disease are marked by an inability to raise the toes while the heel remains on the ground, showing a weakness of the extensors of the toes, and implying a loss of function of the branches of the anterior tibial nerve. It is a curious fact that the muscles supplied by the anterior tibial nerves are not only most frequently affected in multiple neuritis, but they also innervate the region which is frequently involved in poliomyelitis. In the upper extremities the extensors in the forearm are affected in the earliest stages of the disease, and from these parts the paralysis may extend upward in both extremities until the greater number of the muscles are affected or until total paralysis of all the extremities is established. The thenar and hypothenar groups are not affected at the outset. It

will be seen from this that the musculo-spiral nerve in the upper extremity, and its homologue, the peroneal nerve, in the lower extremity, are the nerves first attacked by the disease in a large number of the cases. This peculiar predisposition of these nerves to the disease cannot be explained on the ground of anatomical position, and for the present is as inexplicable as are the facts of elective affinities in so many other diseases.

In some cases the nerves supplying the trunk muscles, including the diaphragm, may be affected, but this is, after all, a very great rarity. A paralysis of the vocal cords due to multiple neuritis is rare enough, as is also an involvement of the facial nerve and of the hypoglossal. I have, however, seen a number of cases, particularly after diphtheria, in which the ocular muscles, the vocal cords, and the muscles of deglutition were paralyzed.

The paralysis, although it may involve a number of groups of muscles, does not affect all equally; those less affected may undergo contracture, such contractures occurring quite as often in the lower limbs as in the upper extremities. Deformities quite as serious and quite as persistent as those in poliomyelitis may therefore result from a chronic multiple neuritis.

As regards the sensory symptoms it is important to note that every form of sensation is about equally impaired; touch, pain, temperature, and muscular sense may suffer equally except in the earlier stages, in which a hyperæsthesia is a more common than hypæsthesia. In a few instances the pain sensibility may be diminished while the sensation of touch remains, and even if the sensibility to pain is intact the conduction of pain impressions may be very much delayed. While pain and paralysis are the distinguishing symptoms of a multiple neuritis it should be remembered that in not a few instances one or the other may be absent, and in some cases pain may never have been present to any marked degree, or may have lasted for so short a time as compared with the paralysis that the patient who is examined at a later stage of the disease fails to remember that pain was ever present. The electrical reactions may exhibit every form of change from a very slight diminution of faradic and galvanic response to a

partial or complete reaction of degeneration. Atrophy is also a prominent and early symptom. The entire absence of bladder and rectum symptoms is of great importance in the diagnosis of multiple neuritis. Exceptions to this rule have been recorded by Oppenheim.

Vasomotor changes in the paralyzed parts constitute a frequent symptom of multiple neuritis. Œdema and glossiness of the skin, which are observed in the majority of the cases of peripheral nerve disease, are present in many cases of multiple neuritis. If the hand is involved there is a peculiar tapering of the fingers, which is quite characteristic of disease of the peripheral nerves.

The course of the disease will vary very much according to the intensity of the cause. As a rule the symptoms increase during the first four to six weeks and then diminish. While the disease is increasing sensory symptoms are particularly prominent. As soon as the acme of the disease has been reached these symptoms become less troublesome. The motor paralysis is recovered from much less quickly than are the disturbances of sensation. This is due not only to the actual loss of innervating power but to the atrophy of the muscles and to the contractures which have been formed. But even in severe cases a favorable change sets in sooner or later. The atrophy diminishes and the strength of the paralyzed limbs slowly increases until complete recovery is established.

In some instances the onset of the disease is a very sudden one. The paralysis spreads rapidly and may resemble the course of Landry's paralysis; but the fact that in multiple neuritis the trunk muscles generally escape and that the paralysis jumps from the lower extremities to the upper without involving the trunk, will help to differentiate it from an acute ascending palsy. While we have good reason to expect every case of multiple neuritis in a child to lead to recovery, death may result in consequence of complicating conditions, such as paralysis of respiratory muscles, or of the heart from disease of the vagus. Bronchitis and pneumonia are complications very much to be feared in children who have any form of multiple neuritis. Cirrhosis of the liver and gross cerebral disturbances, due to alcoholism, which are common causes of death in cases of multiple neuritis in the adult, do not play a similar *rôle* in children. On the other hand, the involvement of the kidneys in cases of diphtheritic palsy is a frequent cause of death. The psychoses described in connection with the multiple neuritis of adults are not met with in children.

Multiple neuritis may result from a number of different causes: 1. Toxic forms, including those due to metallic and non-metallic poisons. Among the former lead, arsenic, and alcohol are the most potent; and in rare instances mercury, carbonic oxide, and phosphorus may bring on a neuritis. In a very careful article in Keating's "Encyclopædia," Dr. Putnam, of Boston, has discussed the palsies of children due to lead and arsenic. He finds that poisoning by arsenic occurs most frequently through the mixtures used to destroy vermin, such as "Rough on Rats," "Paris Green," "German Fly-paper," and the like. Furthermore, arsenic is contained in many of the coverings used for books, toys, confectionery, labels, etc., as well as for artificial flowers. The same author also believes that wall-paper and some special forms of carpet may be the source of poisoning by arsenic. The arsenic is supposed to be communicated to the child in the form of dust distributed in the currents of air, or in the form of some volatile compound which may be formed in a room which contains arsenious wall-paper.

That some cases occur from the continued administration of arsenic in diseases like chorea there is very little doubt, and it is well to watch children carefully, while they are undergoing this form of treatment.* As for lead, it may enter the system from glazed cards, acid fruits and vegetables put up in tins, also from chocolate and other articles wrapped in tinfoil. Lead also enters largely into the composition of rubber, and has been found in the rubber nipples of nursing-bottles. Various kinds of cakes and candies are treated with lead, and prove a prolific source of poisoning in children; but children as well as adults are exposed to lead-poisoning very much more frequently through drinking-water than in any other way. This is more common in country districts, where lead pipes are connected with pumps, or where lead pipes are used to convey water from wells or springs at varying distances to the house. In the large cities this special source of lead-poisoning is extremely rare.

Alcoholic poisoning in children would naturally be considered a rare condition, and so it fortunately is: but among the ignorant classes of every large population parents will be found who take special delight in the fact that their child can partake of everything, including beer or

* I have seen one case of neuritis in the adult after the use of an arsenic paste over a large denuded surface.

whiskey. A case of alcoholic neuritis in a child five years old was reported years ago from Strümpell's clinic. The child was raised in the saloon of its father, and not only drank considerable wine, but also two quarts of beer, every day. The father is quoted as saying that "the child was always thirsty, and of course we could not give it water."

2. The toxæmic forms of multiple neuritis are far more important—at least in children. Among these we class, first of all, the septic forms of multiple neuritis which may occur in children as in adults after injury of any sort. The invasion of micro-organisms is unquestionably the direct cause of the neuritis.

A similar invasion of specific organisms would account for the neuritis of leprosy and of beriberi, but these are so rare in this country, and particularly rare in children, that we need not make any further mention of them.

Malarial neuritis is of great interest to us, for though it has been described chiefly by authors in Eastern countries, it is not unknown in these parts. I have seen several cases of this description in children who were brought to my clinic from the swampy parts of Long Island.

Dr. Browning, of Brooklyn, has reported several such cases, which were supposed to be cases of poliomyelitis. These cases resemble poliomyelitis in their onset and development, and may be easily mistaken for it. In its general symptoms this form differs but very little from other cases of multiple neuritis except that it is associated much more frequently with the intermittent form of fever, that the disease itself undergoes marked remissions, and that the enlargement of the spleen and examination of the blood are likely to give positive proof of the origin of the neuritis. Recovery is a little more rapid, too, in these cases than in those due to other causes.

Among the toxæmic conditions which give rise to multiple neuritis we must include those blood states which follow upon the various acute infectious diseases (diphtheria, small-pox, typhoid fever, tuberculosis, and also syphilis). As the nerve affection is developed a long time after the initial infection it is probable, in the light of recent bacteriological

researches, that some chemical product of the micro-organisms, and not the bacteria themselves, are the direct cause of the neuritis. With the exception of diptheritic multiple neuritis (p. 240) all the forms just mentioned are comparatively rare in children.

In obedience to the ordinary belief, we must concede the existence of the rheumatic forms of multiple neuritis; but I am free to confess that I have seen only a very few cases of multiple neuritis in children which could with some degree of probability be referred to rheumatic causes. In cases of articular rheumatism in children the nerves are sometimes involved, as is proved by pains radiating down the limb through special nerve-trunks. Refrigeration neuritis occurs in children. In one instance this was due to ice-cold baths which a mother gave to her child daily all the year around. Now and then cases come under one's notice of severe forms of polyneuritis in which the true cause cannot be ascertained. Specific and tuberculous forms of multiple neuritis are rare indeed.

PATHOLOGICAL ANATOMY.—It is natural to infer that the changes are very much the same as those to be found in cases of ordinary simple neuritis (see page 209). In multiple neuritis the changes are chiefly of a parenchymatous order, or interstitial in character; the perineurium is not often involved. In some cases, however, the nerve is swollen, the nerve-sheath is hyperæmic, of a deep purplish color, and, according to Eichhorst, may be covered with minute hemorrhages. There may be some slight changes also in the consistence of the nerve. All these conditions, and the appearances of degeneration, will vary according to the stage of the disease at the time of examination. In the earlier stages we often find the nuclei of the fibres enlarged, a proliferation of the connective tissue between the nerve-elements, and numerous round or spindle-shaped cells with myeline granules near the sheath of the nerve. If the specimen is examined at a later period of the disease the acute condition will have passed and the nerve presents the simple appearance of subacute or chronic degeneration of nerve-fibres. (Fig. 57.)

In some cases the muscular tissue is slightly involved,

the fibres are smaller and paler, the transverse striation is less distinct than under normal conditions, while the nuclei of the fibres and of the interstitial tissues may be increased. The changes in the muscle may be both parenchymatous and interstitial.

In considering the pathology of multiple neuritis, the most striking fact is the peculiar action of toxic agents upon the peripheral nerves, without affecting the spinal centres. The farther away the nerve-fibre is from the mother-cell the less able it is to resist morbid agencies; for this reason the more distal parts of the peripheral nerves are affected at an earlier period in multiple neuritis than are the parts of the nerve nearer the spinal centres.

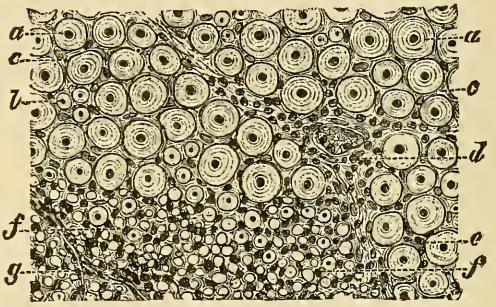


FIG. 57.—Chronic Interstitial Neuritis with Atrophy of some Nerve-fibres. (Ziegler.) *a*, cross-sections of normal nerve-fibres; *b*, cross-section of thin, but normal, nerve-fibres; *c*, endoneurium; *d*, cellular infiltration and proliferation of connective-tissue strands and blood-vessels; *f*, thickened endoneurium with small empty nerve-spaces and some normal thin nerve-fibres; *g*, longitudinal section of a blood-vessel.

This argument is strengthened by the recent conceptions of the close relationship between the ganglion-cell and the peripheral nerve-fibre as parts of a neuron. Moreover, recent researches have shown that the same poison which often affects the peripheral nerve-fibre may occasionally involve the ganglion-cell. Stieglitz has proved this to be the case in lead-poisoning. He found a distinct poliomyelitis in animals compelled to inhale lead salts. Oppenheim has described slight changes in the spinal cord in cases of alcoholic neuritis. Recent investigators have found changes in the spinal and cranial nerve roots, and even in the nerve elements of the brain (Marinesco, Schlesinger). It is certain that in man lead may cause disease of the peripheral nerves.

DIAGNOSIS.—With our present knowledge of the characteristic symptoms of multiple neuritis there should be no difficulty in differentiating between the various forms of this disease and other diseases with somewhat similar symptoms. The chief characteristic of these peripheral nerve diseases is the association of sensory with motor

symptoms, the distribution of both pointing to an involvement of the same nerve-areas. Persistent sensitiveness of the nerve-tracts and subjective pain in the course of the peripheral nerves help to corroborate the diagnosis. It is easy to mistake multiple neuritis for poliomyelitis, and until recent years this mistake was made by the ablest diagnosticians; but in poliomyelitis the initial symptoms are of a rather more violent character, and the entire central nervous system often shows the influences of the disease very much more than it does in multiple neuritis; the paralysis is more symmetrical in neuritis than in poliomyelitis.

In poliomyelitis there is no pain along the nerve-tracts and these nerve-tracts are not sensitive, but vague pains are very often present in the first week or two of poliomyelitis. I have the record of a child, two years old, whom I saw two days after the onset of the paralysis, in whom the entire limb was so painful that the child would scream violently if the limb was suddenly touched, and this was not due to the fear of examination. The pain was not distributed along any nerve-paths, and I did not hesitate to make the diagnosis of poliomyelitis, which proved to be the correct one by the subsequent course of the disease. The electrical reactions are very much the same in both disorders.

The difficulties of this problem are increased by the fact that, as Gowers has reported, cases occur in which a typical anterior poliomyelitis is complicated by a peripheral neuritis. In spite of all efforts at differentiation cases will occur every now and then in which it will be impossible to make an accurate diagnosis at the start, and the physician must watch the course of the disease before coming to a definite conclusion.

From pachymeningitis, which is rare in children, the disease may be differentiated more easily, since in the former the legs are rarely affected, the anæsthesia is apt to spread to the upper limbs and to the trunk of the body, and there is no distinct sensitiveness to pressure on the part of the larger nerve-trunks, except late in the disease, and the atrophy is restricted to muscles innervated from the diseased spinal segment.

Landry's paralysis is to be recognized by the rapidly ascending palsy proceeding from segment to segment, involving the abdominal and thoracic muscles, and not skipping from the lower to the upper extremities, as is the case in poliomyelitis. The differentiation between the ataxic forms of multiple neuritis or the so-called pseudo-tabes from true tabes need hardly be considered in the case of children, as in them true tabes is a disease of exceeding rarity. But it may be well to bear in mind that the symptoms of hereditary ataxia (Friedreich's disease), including the loss of knee-jerk, ataxic gait, weakness, and awkwardness in walking, may bear a superficial resemblance to polyneuritis; but the gradual development of the symptoms in

this hereditary disease, the occurrence in various members of the same family, the peculiar appearance of the face, the disturbance of speech, the mental peculiarities, will render the diagnosis positive enough.

As children are subject to hysterical palsies the question of differential diagnosis may occasionally arise, but hysterical palsy is not characterized by the loss of reflexes; nor is the ataxia, if present, like that of polyneuritis; and, moreover, the anæsthesia accompanying the palsy in hysterical cases is of the regional order, and not in keeping with the anatomical distribution of the nerves. If there is any reason to suspect myelitis, the presence or absence of bladder and rectal symptoms, the development or non-development of bed-sores will weigh heavily for or against the diagnosis of cord disease.

Trichinosis must be suspected in cases of great painfulness of the extremities, but the other attendant symptoms (gastric disturbances) and the excessive swelling of the muscles will help to clear up the diagnosis.

The recognition of the cause of multiple neuritis is an important point in the diagnosis of the disease. In this country the disease in children is developed most frequently after an acute infection, either by poisoning or by the toxic products of some acute infectious disease, such as diphtheria, variola, and typhoid fever. Next in frequency I would place the probability of a malarial origin, and last, the possibility of tubercular, syphilitic, or alcoholic infection.

TREATMENT.—First of all we must determine the primary cause of the disease, and, if possible, the deleterious agent must be removed or its influence arrested. If the child lives in a malarial district it should be taken to a place where there is no danger from further malarial infection. If there is any suspicion of alcoholic, or of lead, or of arsenical poisoning, the sources from which such poisons have emanated must be absolutely cut off. If some acute infectious disease has preceded, we can, of course, do nothing to prevent the natural course of the neuritis, but by improving the child's general health will enable it to conquer the disease much more rapidly. For the relief of pain, which is by far the most annoying symptom at the beginning of the disease, the application of heat is the best remedy we have. If heat is applied at later stages of the disease, when considerable anæsthesia may be present, it is of the greatest importance to watch the condition of the skin, or else serious burns and ulcerations may result. It is on this account, also, that the use of the actual cautery and of other forms of counter-irritation is not to be recommended. Prolonged warm baths will often help to alleviate the suffering, and

will contribute largely to the comfort of the patient. If the pain is so intense that it cannot be relieved by these simple measures it will be well to give small doses of morphia, codeia, or rectal injections of chloral hydrate.

There is no drug which has any specific action upon multiple neuritis. I have given the salicylates invariably, but am not convinced that they have done much good. Mercurials have been praised by some, particularly in cases in which the nerve-sheath is involved rather than in the parenchymatous form, yet I have not been able to persuade myself of the benefits to be derived from mercurials, even in many cases of multiple neuritis in the adult; but if there is any good reason for the employment of mercury let it be given in the form of inunctions of unguentum hydrargyri or of a ten or twelve per cent. oleate of mercury.

The administration of mercury, either by the mouth or in the form of hypodermic injections, is to be avoided in children, if possible. Iodide of potash or iodide of sodium has little or no influence upon the disease; arsenic, which has gained such popularity in every form of chronic nerve disease, is scarcely a safe remedy to use, for in small doses it will have no effect, and if given in large doses there is a decided danger of increasing the neuritis. We are compelled, therefore, in all cases of multiple neuritis in children, particularly in those occurring after diphtheria and other acute infectious diseases, to resort to general tonic remedies. Among these none is better than cod-liver oil, and next in order, though far inferior to it, are iron, quinine, and strychnia in very small doses. In all cases of malarial neuritis, quinine should be given in the same doses that would be administered to combat other symptoms of malarial poisoning. A word of caution should be added with reference to the use of anodynes. In children the careless exhibition of morphia may result in the formation of the morphia habit, and morphia itself, if given in any considerable quantities, may help to intensify rather than to diminish the symptoms.

If insomnia is persistent in consequence of the pain in the initial stages, one of the many hypnotics, such as chloralamid, trional, or veronal, in doses varying between five

and fifteen grains, according to the age of the child, should be given a trial; if they do not produce sleep, codeia in doses of one-sixth to one-half of a grain may be administered.

After the first two or three weeks of the disease the paralytic symptoms become rather distinct. In all those cases in which there is considerable paralysis with atrophy the use of the galvanic current, both as a sedative and as a means of exercising the paralyzed parts should be employed. The faradic current will be of little service if it fails to produce contractions, and as it has a distinct irritating effect upon the sensory filaments in the skin it should not be applied. When all pain has disappeared and the progress of the disease has been stayed, light massage can be employed to advantage; it is specially to be recommended at the time when contractures are about to be developed, or when the atrophy of the muscles is on the increase. If permanent contractures result from multiple neuritis surgical interference may be necessary, but as the disease generally leads to spontaneous recovery, every possible means of bringing this about should be employed before the child is given over to the surgeon; yet I can see no harm in having a simple tenotomy performed if that will enable the child to bring its feet to the ground, or to walk before complete recovery from all the symptoms has set in. In some of the cases some simple orthopedic apparatus will help the child to learn to walk, and will enable it to use muscles which cannot be depended upon to support the body; but there is also a danger in allowing children to wear an apparatus too long a time, as they are very prone to depend entirely upon such artificial support, and, as a rule, lack the energy to exercise weakened muscles.

DIPHTHERITIC PARALYSIS.

Paralysis after diphtheria is far less frequent in this city than it was before the use of the Diphtheria Anti-toxin, and less frequent (relatively) in children than in the adult. In earliest infancy, say up to the age of two years, diphtheritic palsy is extremely rare. It bears no absolute

relation to the severity of the diphtheritic infection. I have often seen typical diphtheritic palsy set in after an infection so slight that it was considered to be nothing but a mild sore throat; but, on the other hand, it also occurs in connection with severe diphtheritic infection, and is frequently associated with other complicating diseases, such as nephritis and endocarditis, following upon the original disease. The previous health of the child has no distinct bearing upon the development of the palsy, strong and weak children being affected with equal frequency. I have seen the paralysis developed within the first week, while the throat was still covered with diphtheritic membrane, but the majority of cases do not occur until two, three, or four weeks after the termination of the disease.

Diphtheritic palsy differs from other forms of multiple neuritis in the order in which the various parts become paralyzed. The palate is generally the first, often the only, part affected. Nasal speech and regurgitation of liquids through the nose are evidences of paralysis of this part, and the loss of function can be determined by an inspection of the mouth and throat. Loss of accommodation is next most frequent. If the paralysis extends further, the upper and lower extremities are apt to be involved. At first a mere weakness is noticeable, associated with tingling and numbness of the parts; later on this weakness increases, and develops into a full-fledged paralysis, and the disturbance of sensation may become more marked. The ocular muscles show a peculiar predisposition to diphtheritic infection; the rectus externus, supplied by the sixth nerve, is frequently, and often singly, affected, and the oculo-motor nerves are also involved at times, but a complete ocular motor palsy in diphtheria is rare. Ptosis and paralysis of one or more muscles supplied by the third nerve have frequently been noticed. The optic nerve fortunately escapes, so that blindness is not a common result of diphtheria, unless by some untoward accident the diphtheritic poison should cause ulceration or purulent disease of the eye, with subsequent loss of vision. The ciliary muscle is often paralyzed; the reaction of the pupils to light is sluggish, but the contraction during accommodation may be preserved,

although the act of accommodation itself is carried out imperfectly.

Paralysis of the pharynx may occur, but is not frequent; the larynx, however, often comes in for a share of the paralysis; the epiglottis then fails to perform its function, and food may reach the larynx and cause severe fits of coughing. Hoarseness and imperfect phonation point to an involvement of the muscles of the larynx and of the vocal cords. This special order of paralysis is generally symmetrical.

The reflexes are diminished or lost, even when no distinct paralysis of the adjacent parts exists or existed (Bernhardt). I have had opportunity to examine several children whose knee-jerks were absent after diphtheria, but who had never presented any paralytic symptoms. The same condition is found occasionally after scarlatina and typhoid fever.

In the limbs the change in the motor and sensory functions is very like that of other cases of multiple neuritis, and does not, therefore, require any special mention. It is questionable whether cardiac failure coming on after diphtheria is due to a neuritis of the pneumogastric, or whether, as Leyden would have it, it is due to a degeneration of the heart muscle itself. The irregularities of respiration associated with the heart symptoms are so characteristic of vagus affections that it seems much more probable to attribute the loss of function to disease of the nerve rather than of the muscular tissue.

Some diphtheritic palsies occur which do not in any way resemble multiple neuritis; a hemiplegia may follow upon diphtheria, but this may be the result of a vascular lesion so common in connection with other infectious diseases of childhood. Cases of this character have been described by Mendel and Bonath.

Paralysis of the masseters has been reported by Grant and quoted by Gowers. As the paralysis was permanent fourteen months after the disease Gowers thinks the affection due to an acute nuclear inflammation. A degeneration of various cranial nerves has been observed to come on after a considerable lapse of time, following upon diphtheria, but it has seemed a little doubtful whether such paralysis could be traced directly to a diphtheritic poison.

The general course of the disease leads to recovery. It may last from six to eight weeks; the cases in which single nerves only are affected recover more rapidly than those with multiple nerve-lesions. The ocular nerves recover

very much more quickly than the nerves of the lower extremities do. If there is paralysis of the legs, it takes, as a rule, from four to six months before the symptoms disappear. The reflexes are late in returning, and in most cases cannot be elicited until some weeks after all other symptoms have passed away.

If death ensues during diphtheritic palsy, it is either due to some complicating disease, such as nephritis or pneumonia, or to paralysis of the heart or respiratory muscles. Mere exhaustion from inability to take food in cases of paralysis of the pharynx is an occasional but rare cause of death.

PATHOLOGICAL ANATOMY.—The morbid changes are essentially the same as those found in other forms of multiple neuritis; the inflammatory and degenerative changes in the nerves are in these cases probably the result of microbic poisoning, or the effect of chemical poisons formed by the diphtheritic micro-organisms; but bacteriological researches have not yet yielded a very satisfactory explanation of all the changes that take place in the peripheral nerves. The rod-shaped bacteria, the bacilli of Klebs and Loeffler, have been found in the blood-vessels of the nerve-centres, and other micrococci have been described by Oertel as occurring in these vessels; but further study will be needed to explain the origin of the inflammatory process in the nerves. The degeneration is found in the nerve supplying the paralyzed part; often the entire nerve is affected, at times some of the filaments only. In others, the degeneration extends to the anterior or even the posterior roots. The myelin and nerve-filaments undergo segmentation, the nuclei of the sheath are increased, and granular corpuscles mark the decay of nerve-tissue. The axis-cylinder remains intact until the destruction of the medullary sheath has advanced considerably, and both these parts may then be entirely destroyed. The interstitial tissue between the nerve-fibres is but little affected, thus marking the condition as a true parenchymatous degeneration. The nerves supplying the palate are most frequently and most severely affected, but the same changes may occur in the peripheral nerves of the extremities in all the ocular nerves, and even the phrenic nerve may be similarly affected.

In diphtheritic paralysis the muscular tissue is more distinctly changed than in other forms of paralysis due to multiple neuritis. In cases of long standing the muscular fibre of the palate, for instance, is found to have undergone granular and fatty degeneration. In some there are signs of parenchymatous as well as of interstitial inflammation, and every possible grade may be found between a simple inflammatory condition and complete degeneration of the muscular fibres. In cases in which the disease leads to implication of the heart muscle, the pallor of the heart after death is the external evidence of complete degeneration of its muscular fibres.

Much has been made of changes in the spinal cord in cases of diphtheritic paralysis, but these are restricted entirely to changes of the ganglion cells of the anterior horns, which have been found swollen, homogeneous in character, and with processes somewhat altered or entirely shrunken ; but it is doubtful whether these changes are primary or secondary, though there is no good reason why a poison which acts so vigorously upon the peripheral nerves should not invade the various tracts of the spinal cord.

The frequent disease of the palate is probably due to the proximity of the nerves supplying the palate, to the diphtheritic membranes, and the easier invasion into these nerves of the micro-organisms lodged in the tonsils, or in the back of the throat. And yet if we remember that a considerable period of time may elapse between the deposit of the diphtheritic membranes in the throat and the appearance of the palsy, and that the palsy may appear after very mild throat affections, it is not altogether easy to explain why these nerves should be so frequently the seat of the disturbance, unless we suppose that from their proximity to the original diphtheritic lesion the nerve-tissue has become altered and thus rendered more susceptible to the diphtheritic microbes, or to the microbic toxins. The poison attacks other nerves in the course of time; the peroneus, the musculo-spiral, and the median are the most susceptible to it.

The diagnosis of this palsy often depends upon the history of preceding diphtheria, or of some throat affection, however slight it may have been. The beginning of the palsy in the palate, and gradually spreading to the lower and upper extremities, will at once suggest the probability of diphtheritic paralysis. It is only in cases in which the original disease has been overlooked or forgotten that an examination of the patient some weeks after the onset of the trouble may, through the atrophic paralysis and the absence of knee-jerk, suggest spinal infantile palsy ; but a closer examination of the patient, and the comparatively rapid development of the palsy, with its tendency to recovery, will place the diagnosis beyond doubt. Diphtheritic palsy, like other forms of paralysis in children, may con-

tinue for an inordinate length of time, and may be superseded by an hysterical palsy. Thus it happens not infrequently, as in the daughter of a clergyman whom I examined many years ago, that a chronic paralysis is the outcome of a diphtheritic palsy which had set in a few weeks after a severe throat affection.

Bear in mind the possibility of cardiac or renal complications. The paralysis of the palate, however disagreeable it may be to the child, rarely leads to serious complications, but if the muscles of the pharynx are involved there is danger of food entering the respiratory tract, and of a subsequent pneumonia. A child seen recently in consultation was in danger of death from this cause.

Other things being equal the earlier the paralysis appears after the initial disease, and the more quickly it reaches an extreme development, the more serious the case and the more imminent the danger to life. It is difficult to forecast the exact length of time which will elapse before complete recovery sets in, but it is safe on the whole to be guided by the same rules as in the case of any other peripheral nerve palsy. If the electrical examination shows an entire loss of response to the faradic current with an altered response to the galvanic current, many weeks will in all probability elapse before recovery sets in. If there are no marked changes in the electrical conditions of the nerves and muscles the degeneration or the inflammation of the nerve is very slight and the paralysis will be recovered from more quickly.

TREATMENT.—The majority of cases of diphtheritic palsy will get well without any treatment, but it is of the utmost importance at all times to maintain the strength of the patient at par, and to be prepared for any sudden complications. If the palate is paralyzed and there is regurgitation of food through the nose it will be better to feed the child by solids than by liquids; if in consequence of paralysis of the pharynx and of the upper air-passages there is danger of food entering the respiratory tract it may be best to feed the child for a time per rectum, or else to use the feeding-tube through the nose or the mouth. In the case of children weakened by the diphtheritic process there is danger of death from exhaustion, and no time should be lost in employing this method of feeding rather than to starve the child while hoping for a spontaneous recovery. On the other hand, even while the rectal feeding is kept up

efforts should be made every now and then to have the child take its food in the natural way. If in any case of diphtheritic palsy there is the slightest irregularity or weakness of the pulse, cardiac stimulants should be given, above all digitalis and small doses of strychnia. If respiratory weakness threatens, the prompt use of the faradic current, as advised by Duchenne, is quite in order. Direct excitation of the phrenic nerves by a slowly interrupted faradic current will have the best results. The electrical currents should be employed, according to the principles laid down in other chapters, for the treatment of paralyzed parts.

The usual tonics—iron, quinine, arsenic, digitalis, and strychnia—should be given, but no one would venture to assert that any one of these has any special curative action except in the way of keeping up the general condition. Indeed there is no better method of treating diphtheritic palsy than by carefully administering food, and making sure that everything taken into the system will help to improve the nutrition of the child.*

LEAD PARALYSIS.

A few pages are to be devoted to this special form of paralysis, in order to emphasize the fact, which was proved conclusively by Dr. Putnam, that children are no less susceptible than adults to the effects of lead-poisoning. Dr. Putnam has not only shown that entire families have been poisoned by lead, but in a series of examinations, made for the purpose of determining the presence of lead in the urine of persons who were supposed to be healthy, he found considerable quantities in the urine of father, mother, and two of the children of one family, all of them being in fair health; a third child, however, for some unknown reason, did not exhibit any traces of the poison. The water drank by this family ran for some distance through lead pipes and was found to contain a large quantity of lead. If present in small quantities, lead is practically harmless. Knowing the varied susceptibility of different persons to lead, and having proved its presence in the urine of persons who exhibited no symptoms of lead-poisoning, Dr. Putnam is right in urging that the possibility of lead-poisoning should be taken into consideration in all obscure cases.

As for the symptoms of lead-poisoning in children, they differ but little from those presented by adults, and are in entire keeping with the symptoms

* Since the above was written, Behring's important discoveries have been made known. Whether diphtheritic palsy can be affected by injections of the serum remains to be seen; but it is my impression that paralytic symptoms have become less frequent. Oppenheim does not share this view.

of multiple neuritis due to other causes. The paralysis is most frequently developed in the extensor groups of the forearm, resulting in characteristic drop-wrist (Fig. 56). The muscles affected are supplied chiefly by the musculo-spiral nerve; the supinator longus, and the extensor of the thumb usually escaping. This escape of the supinator longus is frequently of service in helping to distinguish the disease from a simple traumatic musculo-spiral neuritis, for in the latter case all the muscles supplied by this nerve are about equally affected, whereas lead, like other toxic agents, such as alcohol and arsenic, shows a selective preference for groups of muscles that are functionally, not structurally, related. In adults it is rare to find other muscles seriously affected, except those of the upper extremities, although if careful examination is made of such patients, a slight weakness in the lower extremities and a diminution of the reflexes will be found to be present. I have myself seen two cases of lead palsy in the adult in which the entire affection was restricted to the lower extremities. In one case the adductor muscles of the thighs were the only ones affected. In children the involvement of the lower extremities is not exceptional, and when it does occur, is just as apt as not to occur earliest in the course of the disease, the upper extremities being affected after the lower extremities have been paretic or paralyzed.

While the symptoms are practically identical with those of the adult, the diagnosis of lead paralysis will be all the more readily impressed upon the physician by the occurrence of other symptoms. Thus the child is apt to exhibit a general cachectic appearance; also a peculiar ashen hue of the skin, severe headaches, and marked disturbances of digestion. Soltmann expressed the opinion some years ago that lead colic was rare in children. It is more correct to assert that it is difficult to distinguish lead colic from other forms of colic so frequent in children. Disturbance of digestion due to constipation is the rule, but constipation may alternate with diarrhoea and with severe attacks of colic; the colic being distributed over the entire abdomen and not in the vicinity of the umbilicus, as is generally the case in the adult. Vomiting frequently occurs, and the vomitus may be yellowish in color. The abdomen may be retracted, but in many of the cases one or all of these symptoms may be absent.

If the lead poison attacks the brain, the tendency to convulsions will be very pronounced, and convulsions may possibly be the cause of death. In the cases reported by Karsch and Stewart (quoted by Putnam), the children died in convulsions, one of them with a mild form of delirium. Gowers supposes that epileptic attacks may occasionally originate in lead-poisoning, but Putnam, whose experience in this matter is greater than that of any other author, is inclined to think that such a sequence of events has not been firmly proven.

The lead line, which is of such assistance to us in diagnosticating lead-poisoning in the adult, is not so frequently observed in children, although in one epidemic forty-two cases of the sort were reported. The better condition of the teeth in children is unquestionably the explanation of this.

The pathology and pathological anatomy of lead-poisoning are still subjects for discussion. That lead acts most powerfully on the peripheral nerves cannot be doubted, and that in the majority of cases lead paralysis is the ex-

pression of a neuritis it is also safe to assume. Gombault insists that lead causes a periaxillary neuritis, the sheath of the nerve being more affected than the axis-cylinder and relatively healthy portions of nerve intervening between diseased parts. These changes were found in guinea-pigs that were not paralyzed, and Putnam suggests very correctly that such changes in the myelin without paralysis may account for the fact that in man the electrical reactions are very often altered before any actual paralysis appears. Harnack and others have suggested that lead has a direct action upon the muscles rather than upon the nerves, but the experimental researches of Stieglitz, published a few years ago, have brought the entire discussion to a temporary and satisfactory close by showing that lead produces changes in the spinal cord. Onuf, v. Monakow, and others have reported upon lead poliomyelitis.

PROGNOSIS.—The prognosis of lead-poisoning is, as a rule, entirely favorable. There is more danger from the general nutritional disturbances and from the effect of the poison upon the brain, than from the lesion of the peripheral nerves and of the spinal cord, which may have given rise to the paralysis. If the cachectic condition is recovered from it is more than likely that the patient will regain considerable power in the legs in the course of a month or two, but the actual time that elapses before complete recovery sets in may vary as much in lead palsy as it does in other forms of multiple neuritis, and it need not be a matter of surprise if a child poisoned by lead shows the effects of such poisoning for six months, or even a year; but under all conditions it is a source of satisfaction to be able to predict complete recovery eventually.

TREATMENT.—If the patient is in the acute stage of lead-poisoning the same measures should be employed which are advised in other cases of acute metallic intoxication. If the patient is in a subacute or chronic stage, an attempt should be made to eliminate the poison through the kidneys and skin. The iodides have become the classical remedy. It is better to clear the system of the lead gradually than to attempt to eliminate the poison in a very few days. Doses of from five to ten grains of the iodides three times a day will be quite sufficient for all purposes, and these should be administered in some alkaline water which will stimulate the kidneys to greater activity. Warm baths should be given every day, or every other day, in order to help on the same cause.

After everything has been done to eliminate the lead, the attention should be directed entirely to the treatment of the paralyzed parts, and in this respect the use of active massage and electricity, in the manner so often referred to, is advisable. The general condition of the patient will also call for the active exhibition of blood tonics, such as iron, arsenic, and cod-liver oil.

DISEASES OF THE SPINAL CORD.

CHAPTER XIV.

INFANTILE SPINAL PARALYSIS—THE ESSENTIAL PARALYSIS OF CHILDREN—POLIOMYELITIS ANTERIOR ACUTA.

FEW diseases present as many interesting problems as does infantile spinal paralysis. While the diagnosis of the disease is an easy one, there is considerable doubt as to its true etiology; its pathology, which was supposed to be firmly established, is still under discussion, and the treatment of the disease deserves a most careful consideration. A full history of infantile spinal paralysis would convey a very fair idea of many of the important advances made in neurological science within the last thirty years. In no disease can the improved methods of studying the morbid anatomy of the spinal cord be put to better use, and in few diseases can the finer problems of electrical changes, of the localization of spinal functions, be studied to greater advantage than in this acute spinal palsy.

The clinical features of acute anterior poliomyelitis were recognized many years ago by the German orthopedic surgeon Heine, and his description of the disease still holds good, with few exceptions, to the present day. If we add to his description of the disease what has been discovered since his day, regarding the electrical reactions and the behavior of the reflexes, we shall have an entirely satisfactory account of all the symptoms of the disease. But since the days of Heine innumerable French, German, English, and American authors, among whom we may name Duchenne, Charcot, Marie, Erb, Seguin, Strümpell, Seeligmüller, Siemerling, Sinkler, Mary P.-Jacobi, and a host of others, have contributed to our knowledge of this disease. More recent authors, whose elaborate researches have added greatly to the proper conception of the pathology of the disease, will be referred to later on in connection with this special subdivision of the subject.

SYMPTOMS.—Acute anterior poliomyelitis comes on, in the vast majority of the cases, after the fashion of an acute infectious disease. Its onset is signalized by fever, vomiting,

convulsions, and even coma. All these symptoms may last for a few hours, or even a few days, after which time they gradually subside and give way to the paralysis, which is the most permanent feature of the disease. The paralysis is typical of all the palsies which are due to a lesion in the second division of the motor tract.* This is equivalent to saying that the paralysis is of the flaccid order; that it is associated with atrophy; that the electrical reactions in the paralyzed parts are altered, and that the reflexes in the parts affected are diminished or lost. The very suddenness of the onset is extremely characteristic of the disease. There are, as a rule, no prodromata, and even in cases in which such prodromal symptoms have been reported, it is doubtful whether their occurrence was not a mere coincidence. The fever varies between 102° and 104° F., rarely exceeding the latter for any considerable period of time. The fever generally lasts for twenty-four or forty-eight hours, though in some cases in which

FIG. 58.—Case of Infantile Spinal Palsy; Paralysis and Atrophy of Left Leg chiefly.

*This includes the ganglion cell in the anterior horns, the anterior roots, the peripheral nerve, and the muscle.

all the symptoms show marked severity the fever may last for an entire week. There is no doubt, however, that in a few cases of typical spinal infantile paralysis no fever whatever occurs. Seguin was inclined to doubt this point, but the general consensus of opinion is in favor of the occurrence of such an apyretic condition. The vomiting which accompanies the fever, and sets in at a very early period of the disease, may resemble the cerebral type, and is independent of any gastric disturbance. The convulsions are quite common during the first day of the disease, are occasionally repeated during the first three days, sometimes during the first week. They are of the order of general convulsions, and in this respect can be distinguished from the convulsions which occur during the earlier stage of acute cerebral diseases. In some cases of poliomyelitis anterior acuta the convulsions are entirely absent. Coma is on the whole very much rarer than convulsions, and, if present, may be taken to indicate a tolerably severe form of the disease. The coma is not as profound, and not of as long duration as in many acute cerebral diseases.

FIG. 59.—Case of Acute Infantile *Cerebral* Palsy for Comparison with Fig. 58.

I have drawn this analogy between the acute cerebral and the acute spinal disease, because as a matter of fact a sharp differentiation between the two is often impossible in the earlier days of the disease; and many a case of incipient acute anterior poliomyelitis has been diagnosticated by skilful physicians as the first stage of a meningitis, of cerebral hemorrhage, and what not.

If these symptoms of the initial stage are not well developed it stands to reason that but few physicians would

be willing to make the diagnosis of a spinal infantile palsy with absolute certainty. The diagnosis becomes positive only after the recognition of the form of palsy present.

The paralysis may possibly be present from the very start, but it is very often overlooked in the presence of the other symptoms which appear to be so much more serious. I have not infrequently diagnosticated this spinal paralysis during the first ten hours after the onset of the first symptoms. The paralysis is widely distributed at the start. It may involve all the four extremities, and may even involve parts supplied by the lower cranial nerves (bulbar paralysis) as observed by Medin. In some few cases disturbances of speech (dysarthria rather than aphasia) have been noticed. But the initial wide-spread paralysis rapidly diminishes within a few days, or within the first week, and before long the physician will be able to recognize the parts which will remain permanently paralyzed, or at least to recognize those parts which will remain permanently more affected than others; for it is not until after the lapse of some considerable time, say from two to four weeks, that the retrogression of the paralysis ceases and an inference can be drawn as to the parts that will be permanently maimed. This retrogression of the paralysis has been insisted on by many authors as the most important feature of the early stages of the disease, but it should be added that in some instances the paralysis is seen to increase for a few days after the initial stormy symptoms have passed, then reaches its climax, and from this time on begins to recede until the few groups of muscles that are more seriously diseased give an idea of the amount of permanent injury done.

The permanent paralysis may affect one or more parts of the body, but the lower extremities are much more frequently affected than the upper. In her article in Pepper's "System of Medicine," Mary Putnam-Jacobi, quoting from Duchenne and Seeligmüller, shows that the paralysis was most frequently distributed as follows: Left lower extremity in 34 cases; right lower extremity, 40 times; right upper extremity and left upper extremity, 23 times; all four extremities, 7 times; both upper extremities, 3 times; both lower extremities, 23 times; left upper and lower extremity, twice.; right upper and lower extremity, once; right upper and left lower extremity, 3 times; muscles of trunk and abdomen, once. Of these 137 cases it will be seen that the vast majority involve the lower extremities. The paraplegic and mono-

plegic form are by far the most frequent, while among monoplegias an affection of the lower extremity is more frequent than that of the upper; paralysis of an upper extremity is, however, not so rare but that it is seen often enough in every clinic. Special mention should be made of a paralysis of the deep muscles of the back (which are involved in a number of the cases) which gives rise to extreme lateral curvature of the spine. The hemiplegic variety is very rare indeed.

FIG. 60.—Paralysis of Upper-arm, with Atrophy (Left Side) due to an Attack of Poliomyelitis in Early Childhood.

The further study of the exact distribution of the paralysis in poliomyelitis has brought out a number of interesting facts. E. Remak in particular has shown that the parts paralyzed were functionally, not anatomically, related. In the upper extremity the immunity of the supinator longus in spite of paralysis of all of the extensor muscles in the forearm brings out this peculiarity as well as its affection in connection with paralysis of the deltoid, the biceps, and the brachialis anticus. This latter form of paralysis corresponds with the upper-arm type. In the lower extremity the peroneal group of muscles is more frequently affected than any other; next in frequency the posterior tibial; then the anterior thigh muscles, and least frequently of all, the posterior thigh muscles. The tibialis anticus is generally paralyzed in connection with the quadriceps extensor. These muscles are supplied by different nerves, but are associated in the extension movement of the leg during walking. The involvement of associated muscles in poliomyelitis would furnish the data for the study of spinal localization if the disease led more frequently to a fatal issue and to post-mortem examinations.

An entire extremity, or a large group of muscles, may be permanently paralyzed; in some cases the loss of function may be restricted to a single muscle. There is no little difficulty at times in making out the one or more muscles which have been permanently affected, and in a number of cases

which have come under my observation a difference in the electrical behavior has been the only safe way of determining which muscle showed a departure from the normal.

In addition to this retrogressive form of paralysis, which may at times be widely distributed and at other times curiously limited, we have to notice the rapidly developing atrophy of the paralyzed muscles. I have seen a marked difference between a paralyzed leg and the other normal member within three days of the first onset of the disease, and it is not at all unusual to recognize the wasting of the limb by superficial inspection within the first week or two. The wasting is developed entirely in keeping with the distribution of the paralysis, and this is so true that in one instance, which I can recall, the atrophy as well as the paralysis was evidently restricted to the upper portion of the tibialis anticus. In the case of the deltoid and the trapezius the clavicular portions only may be affected. The majority of the paralyzed limbs present a generally slender appearance. In some instances the difference between the two limbs may not amount to more than an inch or two in circumference, in others the difference may be extreme, and it is not unusual to find a paralyzed extremity that is scarcely half the size of the normal one. The wasted paralyzed part makes the impression of an entirely limp, useless appendage, that is at the mercy of the parts whose muscles are still in a tolerable state of preservation. In addition to the reduction of the muscles the subcutaneous tissue and the fat often disappear, whereas in a few cases of children that were previously well nourished, the muscular wasting may, for a time, be concealed by the very considerable presence of adipose tissue. In addition to the reduction in circumference the atrophied parts do not grow as the normal parts do, whence it follows that short limbs are not infrequently the result of an early attack of poliomyelitis.

The diseased member presents a few other symptoms which render the paralysis easy of detection. The skin over the paralyzed muscles often has a slightly shrivelled appearance; more often still it is blue, cold, and clammy, so that by the mere touch of the two lower extremities the

physician can recognize the one that is paralyzed. This difference in the temperature of the parts is probably due to the improper blood-supply and to the natural shrinkage of blood-vessels that have no duties to perform, or else send a lessened supply to diminutive muscles.

The behavior of the electrical reactions is of the utmost importance. With rare exceptions the paralyzed muscles and the nerves supplying them exhibit a complete reaction of degeneration. Both the nerves and the muscles fail to respond to the faradic current; the nerves cannot be excited by ordinary galvanic stimulation, and the muscle responds in sluggish fashion, and often with an altered galvanic formula, the anodal closure contraction being equal to, or greater than, the cathodal closure contraction (see pp. 44 and 45). These changes in electrical behavior come on very early after the onset of the disease. From several examinations which I have been able to make in very early stages of the disease, I can assert that both the faradic and the galvanic response of the parts paralyzed are increased for the first two days, but then become rapidly diminished; the nerves and muscles soon fail to respond to the faradic current, while the galvanic response may remain increased for a very long period of time; this grows more sluggish, and finally there is no contraction except to very strong currents. It can be stated with some degree of certainty that those parts which continue to respond well to faradism, say after the lapse of a week or more, will not remain permanently paralyzed, whereas the utter loss of function for a varying period of time of those muscles which at once fail to respond to the faradic current may be safely predicted. During the later stages of the disease the return of the faradic response in any muscle, or a normal behavior during galvanic stimulation, will lead us to infer that the muscle, or muscles, in question may recover their previous function, at least to a limited extent; but muscles which exhibit marked electrical changes for a considerable period of time have suffered a very serious injury.

The reflexes are diminished in cases of poliomyelitis anterior; but this is true only of those reflexes which are associated with the normal function of the paralyzed parts.

Thus in a large proportion of the cases of poliomyelitis anterior the knee-jerk is absent; but it is needless to say that it is not absent in cases of the cervical type, nor even in those cases in which only the posterior tibial muscles or the muscles of the feet are involved.

I have in a number of instances been confronted by other physicians with an expression of grave doubt as to the diagnosis of poliomyelitis because of the presence of the knee-jerk, when a careful examination of the parts paralyzed proved that the paralysis was limited to the extensor muscles below the knee and to the posterior groups of muscles, while the quadriceps extensor and all the anterior thigh muscles were entirely normal. The mere presence of the knee-jerk under such conditions need not, therefore, militate against the correctness of the diagnosis.

If all the muscles of a given part are equally paralyzed and equally atrophied no contractures will set in, but all the parts will be equally limp. Thus the leg can be pushed to and fro by the slightest touch (Punchinello leg). Since locomotion and station are the results of an accurate balancing between the extensor and flexor groups of muscles, it is but natural to expect that if one set of muscles is paralyzed, the opposing muscles, being tolerably normal and having less work to do, will overact, and permanent overaction is expressed by contracture. As a rule these contractures do not appear until an attempt is made to use the affected limbs, but in a few instances—and some such have come under my notice—the contractures form while the child lies helpless on its back without any attempt whatever at walking or at using the limb. It would seem from this that some more active cause must be at work, and it is probable that the inflammation which destroys the function of some cells may cause an irritation of neighboring cells and fibres, and thus produce contracture, very much as it it is produced in other diseases in which a lesion in the adjacent white matter acts as an irritant upon the ganglion cells of the anterior horns. According to Volkmann and others the mere weight of the body or of a limb may cause a deformity if the muscles surrounding a joint are paralyzed.

The most frequent deformities are as follows: Pes equinus, or equinovarus; pes valgus; genu recurvatum and incurvatum; permanent flexion (rare); all these deformities are evidently promoted by the attempt to use the

maimed legs. Deformity of the hip is very much rarer than those affecting the knee or the foot. If the disease involve the upper extremities extension of the wrist is common in the presence of paralysis of the flexors; clawed hand if the interossei are involved, and a flexion of the fingers or wrist if the extensors chiefly are paralyzed. The elbow-joint is rarely deformed, but is often entirely useless if the upper-arm muscles are paralyzed. At the shoulder-joint atrophy of the deltoid may be the direct cause of a subluxation. The most serious deformities that occur are those due to a paralysis of the abdominal and deep spinal muscles. According to the extent and number of the muscles involved we may have a scoliosis, occasionally a kyphosis; but most frequently very marked lateral curvature, particularly in the cervical region, or a very marked lordosis in the lumbar region. From paralysis of the abdominal muscles the abdomen may become peculiarly distorted, and in one case of a child of about three, which I saw in private practice, a marked lordosis of the lumbar region was associated with what appeared to be an enormous bulging of the anterior left half of the abdomen. In accounting for these deformities we must remember that in addition to the paralysis of the muscles, the arrested growth of the bones, and the disturbance in the general development of the child play a very considerable part.

Before leaving the symptomatology of the disease it is important to insist on a few negative symptoms which help to corroborate the diagnosis in doubtful cases. It is generally asserted that pain is entirely absent. This is true of the vast majority of cases, and particularly of the later stages of the disease; but in one of the few cases which I was fortunate enough to see at an early day, pain was an extremely prominent symptom. In such cases, therefore, the absence or presence of pain could not serve as a sufficient factor in differentiating the disease from a peripheral neuritis, or from articular rheumatism. The bladder and the rectum are, as a rule, not involved. This is a point of some importance in helping to differentiate between inflammation of the anterior horns and a general myelitis—a point of differential diagnosis that comes up frequently enough for discussion. The entire absence of cerebral symptoms after those of the initial stage have passed away will help to distinguish the spinal infantile paralysis from many cases of cerebral palsy.

Summarizing all these symptoms we may state that the diagnosis of poliomyelitis anterior may be made if the paralysis, however widely distributed, or however narrowly limited, and in whatever part of the body, comes on after

an acute onset marked by fever, vomiting, and convulsions, and if this paralysis is associated at an early day with atrophy, with changes in electrical reactions, and with a loss of reflex activity in the paralyzed parts.

Morbid Anatomy and Pathology. — "Poliomyelitis anterior due to atrophy of the ganglion cells of the anterior horns," has become a byword of neurological science. It would be fortunate indeed if such a simple statement as this would settle the much-disputed question of the morbid changes underlying the disease.

As far back as 1863, Cornil published a case of a woman who had acquired a sudden paralysis of both legs at the age of two years, and who had died of carcinoma at the age of forty-nine. Cornil found the spinal cord much smaller than normal, and thought this due to an atrophy of the anterior roots and of the antero-lateral white matter. He referred to atrophic ganglion cells, but did not make them responsible for the disease. In 1865 Prevost and Vulpian examined another case, also in an elderly person, who had died of cerebro-spinal meningitis. She had had a paralytic club-foot with complete atrophy of the posterior leg muscles, which was found to be due to an atrophy of the anterior horns of the left side, in the lumbar enlargement. The ganglion cells were found to be fewer in number, and the antero-lateral white matter was also atrophied on the same side. Very much the same result was obtained in another case by Clarke, but the importance of disease of the gray matter and of the relations of the ganglion cells of the anterior horns to the paralyzed parts was not fully established until the famous publication of Charcot and Joffroy in 1870, who made the atrophy of the ganglion cells entirely responsible for all the symptoms of the disease. Their studies were based also upon the examination of the spinal cord of a woman, who died at the age of forty, but who had acquired an infantile paralysis at the age of seven years. The ganglion cells in the lumbar segment were diminished, and were in part entirely absent. A dense sclerotic tissue marked the disappearance of these ganglion cells.

Since the publication of Charcot and Joffroy's article, it was current belief that the chief anatomical changes in poliomyelitis were those involving the gray matter and its ganglion cells; that the morbid process might extend throughout the entire length of the cord, or it might be developed chiefly in the cervical or lumbar enlargement. The question arose whether the inflammation of the anterior horns is parenchymatous in character, that is, a primary inflammation of the ganglion cells, or whether it is of the order of interstitial inflammations, in which the neuroglia is the part first affected

and the ganglion cells are destroyed later on. This dispute has been carried on to the present day. It was begun by Roger and Damaschino, and has been continued by Leyden, Schultze, Eisenlohr, Bramwell, Marie, Marinesco and others. Leyden recognized that a number of different pathological processes might give rise to the clinical symptoms of an infantile spinal palsy. He was inclined to adopt Charcot's theory of a parenchymatous inflammation, but in one case which he examined he thought the lesions somewhat similar to those described by Roger and Damaschino. It was evident that this question could not be satisfactorily settled if the microscopical investigations were restricted to cases in which many years had passed between the time of onset of the disease and the death of the patient.

The effort has been made to study recent cases of the disease. The first good opportunity was presented by a very extensive epidemic of poliomyelitis anterior acuta observed by Professor Medin, of Stockholm, and carefully described by his pupil John Rissler, who recorded the autopsies on three cases. Goldscheider, Marie, Redlich, and Siemerling have also published the results of post-mortem examinations of children who died very shortly after the onset of the disease. French and German authors have helped greatly to develop a fuller knowledge of the anatomical process, and their results are in direct contradiction of the opinions of Charcot and Joffroy, which have obtained such general credence in neurology. Nearly all recent writers* have come to the conclusion that the entire gray matter is the seat of interstitial inflammation, and that the changes in the ganglion cells are secondary. Many of the authors are also inclined to regard poliomyelitis as an acute infectious disease, in spite of the entire absence, up to the present day, of proof of the microbic origin of the disease. They are of the opinion that the entire gray substance is easily affected and infected by the poison, and that that part of the spinal cord is most easily involved which has the most abundant blood-supply.

Goldscheider's investigations, which were most carefully made, point to the important *rôle* played by the blood-ves-

* Rissler and v. Kahlden upheld Charcot's views for many years.

sels of the spinal cord. The author concludes that a condition of irritation is present in the walls of the blood-vessels which leads to a dilatation of these vessels and to a proliferation of their endothelial elements. From this the morbid process extends to the neuroglia and produces a proliferation of its cells. The changes in the ganglion cells are of a

FIG. 61.—Poliomyelitis Anterior; Part of an Acute Myelitis. Death of child (age, two and a half years) eight days after onset of complete palsy of legs and arms. (Siemerling.) Section through lumbar segment, showing disruption of anterior gray matter from hemorrhage into it. B, marginal blood-vessel; A, branch of anterior spinal artery.

degenerative nature, and the changes in them, as well as in the nerve-fibres, are secondary and due to disease of the blood-vessels. Goldscheider has also shown that these degenerative changes occur most distinctly in the vicinity of

altered blood-vessels, and that the degenerated ganglion cells lie in vascular areas.

The cases reported by Siemerling, while corroborating the views of Goldscheider and others, have a still deeper significance. They show that in some instances a poliomyelitis anterior is merely a part of a general myelitis of both the gray and the white matter. The frequent restriction of the process to the anterior cornua must be ascribed altogether to the peculiarities of arterial blood supply. It is

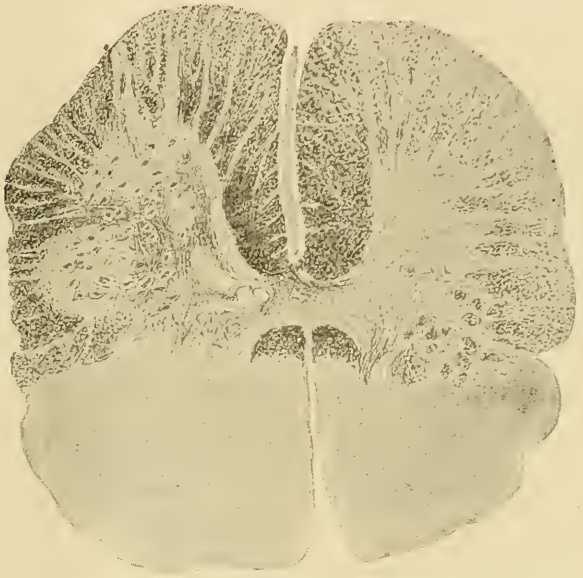


FIG. 62.—Poliomyelitis Anterior of Old Standing, showing Disappearance of Ganglion Cells on Right Side, and Shrinkage of Right Half of Cord. Drawn from a section through lower cervical region.

doubtful, however, whether the hemorrhage into the anterior horns is often as destructive as in one of Siemerling's cases (Fig. 61). All these investigations prove that in infantile spinal palsy the inflammatory process is interstitial not parenchymatous. It may be limited to a few segments of the cord, or it may involve the greater part of the cord, and may extend to the medulla and pons.

For the changes that occur in consequence of this early inflammation, and for the appearance of the cord in the later

years, we may refer safely enough to the famous publications of Charcot and others.

There is, first of all, a disappearance, often complete, of the larger ganglion cells of the anterior horns, and the few that are visible are altered in appearance. The nucleus has disappeared, the cell-body is shrunken, and the cell contours are entirely different from the normal, every trace of the cell processes having disappeared. But the ganglion cells are not the only parts that undergo changes; the entire gray matter shrinks, and, as can be seen in the annexed cut, the entire gray matter in one-half of the spinal cord is shrunken in size, and the white matter of the same side is also very much less in volume than that of the opposite half. In consequence of the changes going on in the gray matter, the columns of Clarke disappear, together with other nervous structures. These changes are unquestionably secondary to the changes in the anterior horns, and considering the intimate relation between the anterior horns and the remaining part of the gray matter, as has been brought to light by recent anatomical investigations, we can readily understand why, in consequence of disease in one part of the gray matter causing destruction of nerve-cells, the nerve-fibres which owe their life and nutrition to such cells disappear as well. The anterior nerve-roots are smaller than the corresponding roots of the sound side. These changes are also, in all probability, secondary to the changes in the ganglion cells.

FIG. 63.—Poliomyelitis Anterior. Chronic stage; section through sixth cervical segment, showing diminution of anterior gray matter and of entire half of right side. (Drawn from a specimen kindly furnished me by Dr. Collins.)

So much for the changes to be observed in the spinal cord itself. The atrophied muscles also present characteristic conditions. The fibres are very much diminished in size, many of them have disappeared altogether, and the place

once occupied by the normal fibres is largely filled by adipose tissue. There is in these cases no such nuclear proliferation and no hypertrophy of fibres such as are found in the muscles of patients suffering from various forms of muscular dystrophy. But even the presence of a few hypertrophied fibres would not be unusual, as the stage of hypertrophy seems to indicate an incipient irritation which precedes the condition of atrophy. Marie has gone to some trouble to show that even the bones in cases of poliomyelitis undergo trophic changes. The bones are smaller than those of the corresponding healthy member and appear more rounded on cross-section than the healthy bone does.

THEORY OF THE DISEASE.—That poliomyelitis represents an acute inflammatory condition of the anterior gray matter of the spinal cord is conceded on all sides, but the question arises what the origin of such inflammation may be. The only satisfactory explanation at the present day is to suppose that the inflammation is the result of an acute infection which happens to be located in the spinal cord, just as other acute infectious diseases show a predilection for other sites in the body. The microbic origin has not yet been satisfactorily demonstrated, but all the clinical facts point toward this view, and the close dependence of the myelitic process upon the distribution of the blood-vessels lends further color to this theory. The infectious origin of poliomyelitis is also rendered highly probable by the frequent observation of the epidemic occurrence of infantile spinal paralysis. Such epidemics have been recorded by Medin and Briegleb in Europe, and by Colmer (1843) and Caverly (1894) in this country. Zappert speaks of a well-nigh epidemic occurrence of the disease in Vienna in 1898.

For several years past I have recorded carefully the cases of poliomyelitis in dispensary and private practice, and have noticed that at least 75 per cent. began between the months of July and October. Medin observed five cases in the spring of one year, and between August and November he had examined altogether forty-four cases of poliomyelitis which had begun during this period. Sinkler states that of 270 cases 213, or 78.8 per cent., were attacked in the hot months of the year, from May to September, inclusive.

Marie supposes that an infectious embolism or thrombosis in one or more of the branches of the anterior spinal

artery may be the direct cause of the attack of poliomyelitis.

Marie is inclined also to infer the infectious nature of poliomyelitis from the close resemblance between polioencephalitis and poliomyelitis, and quotes approvingly the two cases of Moebius occurring in one family, in which one child was attacked with the form of acute cerebral palsy, and the other child with an acute spinal palsy. But surely this proof of the infectious theory of acute cerebral palsy is extremely slender, and Moebius's cases might well be due to a coincidence rather than to an infection, which is supposed to have caused a cerebral paralysis in the one, and a spinal paralysis in the other child. Nor can two cases occurring in the same family be considered evidence of an epidemic character of the disease. A few years ago two children, cousins, were brought to me, who had developed acute spinal palsy within two weeks of one another. The one child has remained severely paralyzed in both lower extremities up to the present day, the other child escaped with a slight paralysis of the anterior tibial group. The theory of infection would be a very simple one to hold in such cases, but on closer examination it was found that both these children had developed the symptoms of their disease shortly after exposure to an extremely cold surf bath. The infectious theory of poliomyelitis is a very plausible one, but we cannot disregard other possible causes, and refrigeration, as in the two cases just cited, may in some instances be a powerful factor in the development of poliomyelitis. It seems to me that those authors make a mistake who insist on a single origin for such a frequent disease as poliomyelitis. In a very recent article (Brain, Autumn, 1904), Batten arrives at the same conclusion: "Such thrombosis (of the anterior spinal artery or its branches) may be produced by many and various forms of infection and the disease is not due to a special specific infection." Schultze reports that he has found Weichselbaum's diplococcus in the cerebro-spinal fluid obtained by lumbar puncture; he thinks that occasionally poliomyelitis anterior may be a disguised form of cerebro-spinal meningitis. Marinesco is of the opinion that the entrance of micro-organisms into the anterior spinal artery produces poliomyelitis anterior. Great caution must still be exercised in the interpretation of bacteriologic findings.

Differential Diagnosis.—Meningitis can be excluded if there are no other signs of a meningeal process, except possibly coma and convulsions. In cases of meningitis these constitute the first of a series of many cerebral symptoms, such as vomiting, rigidity of the neck, headaches, cranial nerve affections, and the like. In poliomyelitis none of these symptoms appear, and the coma and convulsions last but a relatively short time.

There is little difficulty in distinguishing between well-

developed cases of acute spinal and acute cerebral palsy of children, but the less pronounced types of these diseases cannot be easily distinguished from one another unless a very careful examination is made of all the accompanying symptoms. The mode of onset may be exactly similar in both; it is, in fact, on the close resemblance between the two diseases in this respect that Strümpell was led to build up his theory of the analogy between the two. But aside from the symptoms of onset, the clinical features are almost diametrically opposed to one another. The following table will bring out these symptoms in the clearest possible manner:

Acute Spinal Palsy.	Acute Cerebral Palsy.
Onset sudden, with fever, coma, and convulsions. Convulsions rarely repeated after first few days.	Onset sudden, with fever, coma, and convulsions. Convulsions apt to be repeated.
Paralysis flaccid, associated with atrophy.	Paralysis spastic; no atrophy; associated with rigidity and contractures.
Paralysis widely distributed, possibly involving all extremities, or narrowly limited to one member, or even a single group of muscles.	Paralysis generally hemiplegic, sometimes diplegic or paraplegic. Monoplegia rare.
Electrical reactions altered (R. D.).	Electrical reaction normal.
Deep reflexes diminished or lost.	Deep reflexes exaggerated.
Intellect never permanently involved; no epilepsy.	Intellect often involved; epilepsy frequent.

Doubt may arise as to the differential diagnosis in some cases between poliomyelitis anterior and a peripheral (multiple or simple) neuritis. The onset may be equally sudden in both, though in many cases of neuritis the onset is much more gradual than it is in cases of poliomyelitis. In neuritis there are, as a rule, fewer symptoms of general nervous disturbance than in poliomyelitis; but in those forms of neuritis in which there is a distinct toxic infection the toxic poisoning may produce cerebral symptoms very closely akin to those met with in the earlier stages of poliomyelitis. The distribution of the paralysis may be the same in both instances, but after all it is much more likely to be distributed according to strict anatomical lines in

neuritis than in poliomyelitis. In the latter, muscles that have a common function are very apt to be paralyzed together. In former days the presence of pain along nerve-trunks and along nerve-branches supplying the paralyzed muscles was supposed to be a safe feature of differential diagnosis, and this holds good in a majority of cases; but according to my own experience pain may be present in the acute stage of poliomyelitis, and if the child is too young to give accurate information to the physician, it is well-nigh impossible to determine whether the pain is a general one in the joints or whether it is along the distribution of the peripheral nerve-branches. But I have never seen pain persist for any great length of time in poliomyelitis, while it persists, as a rule, for days and weeks in cases of neuritis. The atrophy, the electrical reactions, and the reflexes may be as thoroughly affected in one disease as in the other. The differential diagnosis can, in many instances, be made only after a close observation of the entire course the disease has taken. (See Chapter on Multiple Neuritis.)

The various forms of progressive muscular dystrophies may occasionally be mistaken for poliomyelitis and *vice versa*. In cases that are seen years after the onset of the trouble, the initial history of the case will often be an important guide to diagnosis. If a case of typical muscular dystrophy is seen during the stage of atrophy, a superficial inspection of the case may suggest an old poliomyelitis; but in the progressive dystrophies the atrophy affects an entire limb rather than single groups, or if it has spread to several extremities it is, as a rule, much more general than in cases of poliomyelitis, while in the latter the electrical changes are, as a rule, much more complete than in cases of progressive dystrophies.

The Charcot-Marie type (Tooth, Hoffmann, myself and others) must be differentiated from subacute poliomyelitis.

In the year 1900 Oppenheim described a condition observed in very young children which he designated as "Myatonia congenita." The symptoms bear close resemblance to those of poliomyelitis anterior. The legs and possibly the arms of the child thus afflicted are limp. The trunk muscles are rarely affected. The deep reflexes may be diminished or lost, but the ex-

tremities are not as completely paralyzed or even as paretic as at first sight they would seem to be. The electrical behavior of the muscles may be more or less altered. There is, of course, no history of an acute onset, and the entire state bears the earmarks of a condition of defective development.

I have on many occasions called attention to the muscular weakness associated with rickets and which may easily be confounded with poliomyelitis anterior.

PROGNOSIS.—Altogether too gloomy a prognosis is generally given in poliomyelitis. This is based upon the fact that some palsy always remains, but the actual residue of palsy may be so slight that one should be careful not to depress the hopes of parents and patient. Above all, there is no need of predicting that the child will remain a hopeless cripple for life. There is no telling at the outset of the disease to what extent the retrogression may take place; but, of course, the more widely distributed the paralysis is at the beginning the larger the remaining palsy is apt to be, although some cases which begin in very stormy fashion exhibit more progress than those which begin less violently. Cases in which but a few muscles are paralyzed at the start often recover with very little permanent injury. Very little change need be expected in the first few weeks of the disease, but there is reason to hope that those parts which show any improvement within the first few weeks or months after the onset of the disease will recover power before long, and only those parts will remain permanently paralyzed which after months show no signs of improvement. Muscles which are paralyzed, but which exhibit slight, or no changes of electrical reaction, may be regarded in a hopeful light, and, on the other hand, those which very soon after the onset of the palsy exhibit distinct reaction of degeneration, and for months afterward show no sign of change in this respect, are apt to be permanently paralyzed. The more complete the wasting of the muscles, the less likely these muscles are to recover; and if contractures form in the opposing groups permanent disability is the probable result, but even such disability can often be remedied by surgical procedures.

The prognosis as regards life is, with few exceptions, entirely favorable. Cases that end fatally are apt to do so

within the first few weeks of the disease. But parents are often most grateful for the assurance that if the child survives, however great the paralysis may be, its mental development will in nowise be impaired. Hirsch, Cassirer, and others have shown that individuals who had poliomyelitis may develop amyotrophies or dystrophies.

TREATMENT.—During the acute stage of an anterior poliomyelitis the general condition only should be treated and little attention need be paid to the paralysis. The child should be kept in a quiet room, mild antipyretic measures may be employed, such as small doses of phenacetin, of antipyrin, of the salicylates, and the like. In the earlier stages cold applications, or mild counter-irritation over that part of the spine which is involved in the given case, are quite in order; and the attempt should be made to limit the spread of the inflammation by the administration of small doses of bromide and of ergot (a few drops of the fluid extract). Iodides and other drugs I have found to be utterly inefficient. The child should during this period be carefully fed, and the bowels should be thorougly purged by the use of small but sufficient doses of calomel. After the acute stage is passed the paralyzed muscles demand treatment. Electricity and massage are the most effective therapeutic measures. Avoid electrization of the spine; first, because it is not at all certain that the electric current reaches the spinal cord, and secondly, because the use of strong currents makes the child extremely restless and may do more harm than good. In the treatment of paralyzed muscles an important use of the electric current is to exercise muscles which are no longer subject to the will; it supplies, in other words, a convenient form of gymnastics. The current may, in addition, improve the state of the paralyzed and atrophied muscles, but whether or not it increases the conductive powers of paralyzed nerve and muscle I am not willing to assert. Since we wish to make the muscles contract, the only form of current that is serviceable is that form to which the muscle will respond. If the reaction of degeneration is complete the faradic current is quite useless, and if the diagnostic tests have shown that the muscles respond to the anode better than to the cathode, exercise by

anodal opening or closing of the current is the only proper method.

This should be done in sittings of ten to fifteen minutes once or twice a day, and that strength of current should be employed which is sufficient to produce mild contractions. Excessive contractions are not called for and help to increase the difficulties of application. If a muscle responds to the faradic current, however slightly, that current should be employed together with the galvanic, and it has been my habit, even in the more severely paralyzed cases of poliomyelitis, to make occasional tests with the faradic current, first, in order to determine whether there is any sign of improvement in any group of muscles that have been paralyzed, and, secondly, to give such muscles the benefit of both currents. If the muscles react at all to both currents, both may be employed in one and the same sitting, or given in alternate sittings.* I am a thorough believer in the good influence that massage has upon atrophied and palsied muscles. It helps undoubtedly to keep up the nutrition of such parts, and in cases in which there is an incipient tendency to contracture such tendency may be overcome by the proper use of massage. But this should, if possible, be intrusted to skilled manipulators and not to the mother of the child or to a nurse, whose "rubbings" are, as a rule, wholly ineffective. Passive movements are entirely in order, and some good results have been attained by the regular use of Swedish movements, such as are given by trained rubbers or by a regular system of treatment in a well-equipped Zander institute.

During the chronic stages of poliomyelitis orthopedic measures should be employed without reserve. If the contractures have persisted for years they will not disappear without treatment. Tenotomies are followed by results fully as favorable as those in chronic ocular palsies. The general condition of the child and the condition of the paralyzed muscles improves markedly after the orthopedic surgeon has done his work, and such tenotomies need not be restricted to the tendon Achillis, but many of the other muscles which are in a state of contracture can as well be similarly treated. The application of splints according to the best orthopedic principles is also of great assistance to the child, and this, too, should not be delayed too long, for it is far better to have a child walk in splints than to have ugly deformities of the joints develop which may cripple him for life. Since I have recognized the truth of these principles I have had the satisfaction of seeing children,

* I have seen no reason to advocate static or high tension currents in cases of poliomyelitis.

and even very young children, walk within a few months after the development of a poliomyelitis, while in former years such children were compelled to be carried about in the arms of a nurse or to be wheeled about in chairs. I am satisfied that few cases of poliomyelitis are so severe that much cannot be done by the proper application of orthopedic measures. In cases in which the joints are entirely useless on account of the complete atrophy of the muscles, the operation for arthrodesis, as suggested by Wolff and others, may be resorted to. In this way a leg that would otherwise be entirely useless may be made to subserve the function of standing and walking, though of course the station and the gait of the person will always be far from normal. Many years ago Nicoladoni suggested the transplantation of tendons in these cases. The "method" was revived by Gluck, Lange, Vulpius, and Hoffa. Townsend and Gerster have operated upon a number of my patients. The principle is a simple one: Split the tendon of the overacting muscle, and transplant it into the underacting; but make sure that the "overacting" muscles are in good condition and that the "underacting" muscles are not *completely* wasted. To correct the position of flaccid limbs good can be done by shortening of the tendons as practised by Hoffa. Much is to be expected of the orthopedic surgeon in this field. Spiller, Young, and Frazier have attempted in cases of poliomyelitis in which only a few muscles were paralyzed to improve the condition of the limb by nerve-transplantation. (See Journal of Nervous Mental Disease, 1903, p. 369; also Journal of Am. Med. Ass., 1905, p. 169.)

SUBACUTE ANTERIOR POLIOMYELITIS.

Subacute anterior poliomyelitis is practically a mere variety of the acute form of the disease, and for that reason needs but little special mention. The entire difference between the two diseases is in the mode of onset, and according to our present views of the character and origin of acute spinal palsy, the subacute variety necessarily implies a milder form of infection than in the cases with a more acute and more violent beginning. The difference in the symptoms is also confined entirely to the difference in the manner of onset and the manner in which the paralysis is developed. In these cases of the subacute variety we find that the disease comes on very gradually. The

child is ill at ease for some days or weeks, complains of weariness in walking, of pains in the joints and muscles. After some little while a decided paresis of one or more groups of muscles is observed, generally in the lower extremities. This increases, and after a week or more a distinct paralysis is developed. The paralysis then increases in the affected muscles, and is apt to spread somewhat after the fashion of a progressive muscular atrophy from one group of muscles to another. There is, therefore, a progression in these cases at the start, but the limit is very soon reached, and from this time on a retrogression again sets in, though not in such a marked degree as in the acute cases. The fact that the retrogression occurs after a given period of time will dispel all fears as regards the possibility of a progressive form of muscular atrophy.

The differential diagnosis between these diseases is difficult, and cases are confounded easily with forms of progressive muscular atrophy, particularly of the peroneal type, and with cases of chronic neuritis; but the points of differential diagnosis which were given between these diseases and the acute form of anterior poliomyelitis will also help us in arriving at a correct diagnosis of the subacute variety.

TREATMENT.—The treatment should be conducted on the same principles as were enunciated in the preceding discussion on acute spinal palsy. In those cases in which the original disease is not nearly so violent as in the acute form even more can be hoped from an early application of therapeutic measures, but it should be remembered that the natural course of the disease tends much more to recovery than does the acute form, so that tenotomies and other surgical procedures should be delayed until the disease has become entirely stationary, and there is no reason to think that further spontaneous recovery will take place.

CHAPTER XV.

ACUTE MYELITIS.

MYELITIS, or inflammation of the spinal cord, has been made to cover a multitude of diagnostic sins, both in the adult and in the child. If we subdivide the cases according to the mode of onset, we may distinguish between an acute, a subacute, and a chronic form; and if the classification is based on the origin of the myelitis we have an idiopathic, a traumatic, a tubercular, a syphilitic, and an infectious variety. As the symptoms are very much the same, whatever the original cause of the disease may be, it will be better to describe the characteristic features of acute myelitis, which occurs frequently enough in children to demand special study.

SYMPTOMS.—The symptoms in a given case will vary according to the site of the inflammation, and the intensity of the process. Its clinical features will depend upon the amount of cord tissue involved. In such cases more than in any others an accurate knowledge of the functions connected with each segment of the cord is of importance if a satisfactory diagnosis is to be made. In every case of complete transverse myelitis, at whatever level the area of inflammation may be, motion and sensation are chiefly affected, the reflexes are disturbed, and the functions of the bladder and rectum are deranged. Bilateral paralysis (paraplegia) is the natural result of a myelitis. If the lesion is in the cervical portion of the cord, both upper and both lower extremities will be paralyzed; the first because the very parts which are most intimately connected with the motion of the upper extremities are destroyed; and the lower extremities are involved because the fibres going to them are interrupted at the site of the lesion. The bilateral character

of a palsy, is after all, the one symptom which points more frequently to a spinal lesion than any other.

Almost the only exceptions to this rule are those cases in which a multiple neuritis, an ascending Landry's paralysis, or double cerebral lesions give rise to a bilateral form of palsy.

If the lesion is in the cervical portion of the spinal cord, the paralysis of the upper extremities will be of a flaccid order; the paralysis of the lower extremities will be spastic in character. Anæsthesia will be present in the four extremities and in the trunk to the level of the diseased segments; pupillary symptoms, unilateral blushing (due to lesion of the sympathetic), and paralysis of the diaphragm are present in some cases. If the lesion is in the lumbar portion of the cord, the paralysis is restricted to the lower extremities and will be of a flaccid character, with more or less atrophy. If the lesion is in the cervical or dorsal portion of the cord, and the lumbar portion is entirely free from disease, the paraplegia of the lower extremities is of the spastic order, and the reason of this can be easily understood if we recall the fact that after a transverse lesion in any portion of the spinal cord the lateral columns will degenerate downward from that level (Fig. 64), and that such degeneration of the lateral columns in the presence of normal gray matter of the lumbar segments will produce a spastic form of paralysis with rigidities and contractures. Taking all cases of myelitis, the largest number affect the dorsal region; and in these the arms go free, as their spinal centres are above the site of the lesion, but the lumbar segments are affected by secondary degeneration.

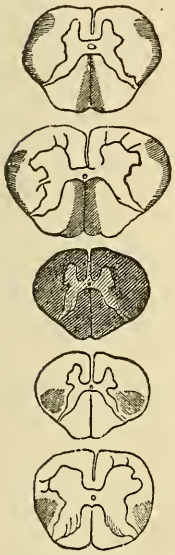


FIG. 64.—Secondary Ascending and Descending Degeneration following a Transverse Lesion in the Upper Dorsal Cord. (Struempell.)

Sensation is impaired at a very early period of the disease, often from the first moment of onset. In fact, in the cases of traumatic origin loss of power and loss of sensa-

tion are effected almost instantaneously, so that a person thus afflicted feels not only that his legs are powerless, but that they appear to be dead as well. Anæsthesia exists in all the parts that are supplied by nerves coming off below the site of the spinal lesion, and in the typical cases all forms of sensation are equally involved. Thus we generally find that a spastic paraplegia is associated with the loss of touch sense, of pain sense, of thermal sense, and of muscular sense in the affected parts. As the anæsthesia is strictly dependent upon the conduction of impulses inward through the posterior roots of the cord, the extent of the anæsthetic area will naturally point to the upper limit of disease.* At the upper limit of the anæsthetic area a small zone of hyperæsthesia, as a rule, begins. This is evidence of the fact that in the segment supplying the hyperæsthetic area there is a condition of irritation affecting the posterior spinal root-fibres, but not the chief morbid and destructive process. Above the hyperæsthetic area sensation may be expected to be entirely normal. In other cases, instead of a distinct hyperæsthesia we have a girdle sensation, which also marks the level between the normal and diseased segments.

The state of the reflexes helps us also to determine the area involved. If the lesion is in the cervical region all the reflexes of the upper extremities are destroyed, those in parts below will be exaggerated. If the lesion is in the dorsal region the reflexes connected with these segments, such as the abdominal and epigastric reflexes, will be lost and the lower reflexes will be increased. If the lesion is in the lumbar region the knee-jerk will be lost and the ankle clonus will be absent also. In some cases in which there is a very narrow band of inflammation these reflexes may behave differently and may give one a direct clew as to the exact extent of spinal inflammation. Thus in one case under my observation the knee-jerk was lost, but the ankle clonus was present. This, taken in conjunction with a line of anæsthesia showing an involvement of the lower dorsal segment, proved that the upper lumbar region was slightly involved, but that the lower lumbar and sacral segments

* The reader is advised to consult special tables on Spinal Localization.

were not directly implicated by the disease. Since dorsal myelitis is the most frequent form, it is also common to have exaggeration of the reflexes associated with spastic contractures of the legs.

The electrical reactions will vary according to the segments involved. In cases of cervical lesion the reaction of degeneration will be found present in many, if not all of the muscles of the upper extremities, at least after the lapse of a few days or weeks; those in the lower extremities will remain unaltered. But if the lesion is in the lumbar segments the reaction of degeneration will be present in the muscles supplied by nerves coming off from the diseased area. In cases of cervical and dorsal myelitis we may, therefore, expect entirely normal electrical reactions in the lower extremities, however thoroughly paralyzed these parts may be.

A few other symptoms must be noted which are extremely characteristic of acute myelitis and often serve to reveal the disease when other symptoms in the case have left the diagnosis in doubt. In all such cases of myelitis, in whatever region the lesion may be, the vesical and rectal reflexes are disturbed or completely abolished. The result of this is retention of urine, with possible overflow, or constant dribbling of urine, and either retention of stool or involuntary defecation. Since the centres for these reflexes are in the lowest portion of the cord, and every part of the cord must be intact if such sensation is to be conducted to the higher centres, we can understand why these symptoms should be present, whether the lesion be in the lumbar, in the dorsal, or in the cervical segments. Loss of sexual function is hardly to be mentioned in cases of myelitis in children; but priapism, due to irritation of the spinal sexual centre, is not infrequently present in children, and is sometimes a very annoying symptom. Involuntary spasmodic twitchings occur in nearly every form of myelitis. I know of no symptom which is on the whole more characteristic of spinal lesions, and which often serves as a differential symptom between supposed functional and spinal paralysis. This spasmodic cramp is evidently due to an irritation of the normal ganglion cells, and the irritability is often so

great that the mildest form of sensory impulse is sufficient to elicit such involuntary spasms. Under these circumstances the mere touch of the paralyzed part—of a toe, for instance—is sufficient to produce contraction of the entire limb.

Trophic disturbances are exceedingly common and much to be feared. Bed-sores are easily developed in all parts on which pressure is exerted: under the shoulder-blades, over the sacrum, on the hips, and even over the internal malleoli or on the inner surfaces of the knees and thighs—in short, wherever parts touch, are pressed upon, or are pressed against each other. In the sacral region the constant wetting of the bed and the uncleanliness of the patient may increase the danger and size of bed-sores; and while it is true that such bed-sores may occur even without any external irritation, they are greatly aggravated by the dribbling of urine or the involuntary evacuation of the bowels. These bed-sores begin, as a rule, as a mere reddening of the skin; the epidermis is soon worn away, the cutis is bared, this too disappears, and gradually the ulcer may eat away all the subjacent parts until the bone itself is laid bare, provided the patient lives a sufficient period of time.

Every form of acute myelitis may be accompanied by fever, which may vary between 100° and 104° F. and higher. The fever is due to the myelitic process, or the infection, but is often increased, and sometimes maintained altogether, by the complicating conditions of myelitis. Such complications are deep bed-sores with the absorption of putrid matter and the danger of phlebitis; furthermore, the occurrence of cystitis and pyelonephritis, which are not uncommon. In cases of myelitis, in which fever suddenly increases, with chills, with deep remissions and sudden exacerbations, the probability of this fever being due to some pyæmic process is very great indeed. As soon as the disease has passed the acute stage the fever lessens and the temperature will remain entirely normal until some complicating condition is established.

Whenever a majority of the above symptoms are present, the diagnosis of an acute (transverse) myelitis can safely

be made. The modification of the symptoms if the myelitis is not complete, or if it is subacute and chronic can be easily inferred from the preceding account. The question of greatest interest, in every case, is to determine what the origin of the myelitis may be. Idiopathic myelitis is, according to our present notions, scarcely conceivable, and here, as in so many other instances, it is better to say myelitis from unknown cause than to assume that in some cases the origin is truly spontaneous. In cases occurring in children, without known cause, I am inclined to suspect slight traumatism.

A child of eight years, a healthy, beautiful girl, while walking on a country road, had a desire to urinate. Her mother urged her to do so on the road. The child hurried to one side, and in attempting to place its right foot on a stone not more than a few inches in height, lost its balance and struck on the middle of the back. The child experienced pain at once, but was able to walk some little distance; soon the power of its legs diminished; it had to be carried, was put to bed, and within twenty-four hours had developed a most pronounced form of acute transverse myelitis. After three days anæsthesia was complete up to the umbilicus. The paralysis was absolute in the lower extremities. There was retention of urine and fæces, bed-sores were developed, and the child died from these complicating conditions within three weeks after the accident.

In other cases the spinal injury is much more severe, and a complete destruction of the cord may be the result of injury to the spinal column. From the effect of concussion alone, without actual destruction of the bony parts surrounding the cord, an acute myelitis may result. The myelitis which occurs in connection with tumors of the cord, with tubercular affections of the meninges, need not be separately considered, as it constitutes merely a part of the more serious disease. The two forms of myelitis which are most common in children are those due to Pott's disease and to syphilitic infection; but both these forms are so distinct and so important that they deserve special consideration.

Acute myelitis does, however, occur in connection with other acute infectious diseases, such as typhoid, scarlet fever, small-pox, and the like. Some of these cases of supposed myelitis have probably been cases of multiple neuritis. Rheumatic or atmospheric influences (refrigera-

tion), as a direct cause of myelitis in the child and in early youth, should be considered duly. A young girl, of about sixteen years of age, was brought to me from the South, with the following history: On a very warm day in early spring she had taken a warm bath and had sat down at an open window in the evening, immediately after the bath, with nothing but a light chemise to cover her body. She sat there for hours and fell soundly asleep. The next morning she experienced considerable difficulty in the use of both upper extremities. Within a few days these became absolutely paralyzed, and the legs at the same time grew stiff and motionless. She has since that time, a period of fully sixteen years, been suffering from the effects of this myelitis, and even now presents an atrophic form of paralysis of the right upper extremity, with slight involvement of the left, and with a complete spastic paralysis of both lower extremities, with incontinence of urine and fæces, and with considerable disturbances of sensation, though the latter have been recovered from very much more than has been the paralysis or the atrophy.

The infectious origin of myelitis has been demonstrated. Streptococci and staphylococci have been found by Marinesco, Tooth, Russell, and others. The author has reported a case occurring in a young girl in association with multiple streptococci abscesses. Fürstner found pneumococci in a myelitic focus. Strümpell lays great stress on the toxic rather than the infectious origin of myelitis.

Pathology and Morbid Anatomy. — The delicate structure of the spinal cord seems peculiarly liable to inflammatory disease. The cervical and lumbar enlargements of the cord are less frequently the seat of such inflammation than the dorsal portion. The reason of this is not easy to explain, although it must in all probability be sought in the peculiarity of the blood-supply.

There can be little doubt that if the blood-supply of the cord is interfered with, necrotic softening follows as in the case of other organs. In the case of one form of myelitis—that due to specific disease—the relation of the myelitis to disease of the blood-vessels can be clearly demonstrated. Under these conditions the smaller vessels are blocked by thrombi, and the result of this obstruction is a necrotic softening of the surrounding parts. In the cases of traumatic myelitis the earliest changes are due to mechanical injury of the part, with compression of the delicate structures of the cord by effusion of

blood, and to the necrosis that follows such compression. It is more difficult to explain the exact manner in which myelitis is developed in the cases of a toxic character, unless we suppose that the chemically altered states of the blood produce coagulation of the blood and obstruction of blood-vessels, with the same result as in those cases in which these conditions are brought about by other disease of the blood-vessels themselves. The origin of myelitis from refrigeration and rheumatic influences in general cannot be satisfactorily explained in this way, nor can any other plausible explanation be substituted. The myelitis which results from mere concussion without any visible anatomical changes must be explained on the supposition that minute changes in the gray and the white matter are present, such as were found in the spinal cords of animals experimented on by Schmauss.

If the exact mode of origin of various forms of myelitis is still unknown, the morbid anatomy is no longer a matter of doubt. As for the macroscopical appearance of myelitis, the cord so diseased is generally surrounded by the hyperæmic meninges, and the cord itself, if inflammation is recent, may appear to be congested and slightly swollen. The distinction between the white and the gray matter is often not so marked as in the normal cord. There is, furthermore, a change in the consistence of the cord, which may be either slightly softer than normal or else so diffluent that as soon as the pia is cut open the cord flows out like creamy pus. This is the condition often found on post-mortem examination; in all probability the cord is not nearly so soft during life, but, like other necrosed tissue, softens considerably immediately after death. In acute myelitis minute hemorrhages are extremely frequent, and the altered cord may present the appearance of red softening. There is every degree of change between simple red softening and the condition of hemorrhagic myelitis in which the extravasation of blood, being considerable, for the time obscures all other changes. If the blood has been exuded for some time before death its color may have changed, and the condition be that of yellow softening. We also may discern a condition of white softening in which the white matter has become diffluent without any admixture of blood. If examined microscopically the cord is found to contain ample evidence of inflammatory changes. Among these are dilated blood-vessels, with leucocytes, granules of myelin, and, furthermore, bodies well known as corpora amylacea. The

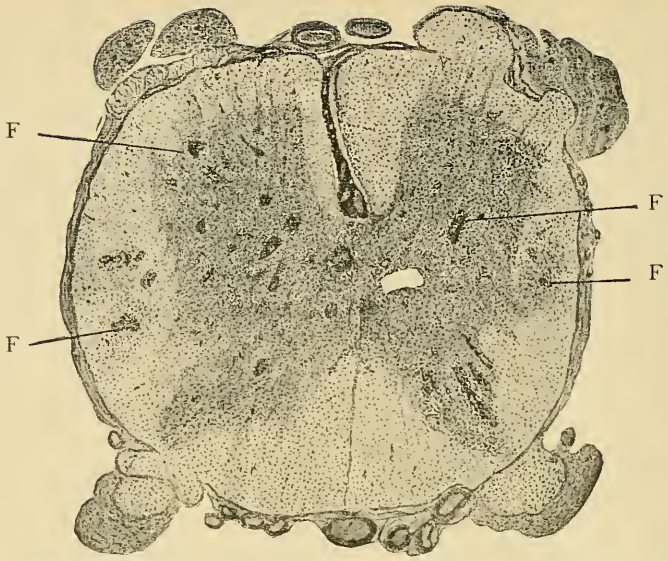


FIG. 65.—Acute Disseminated Myelitis. F, small foci containing round cells generally clustered about blood vessels. van Gieson stain (after Oppenheim).

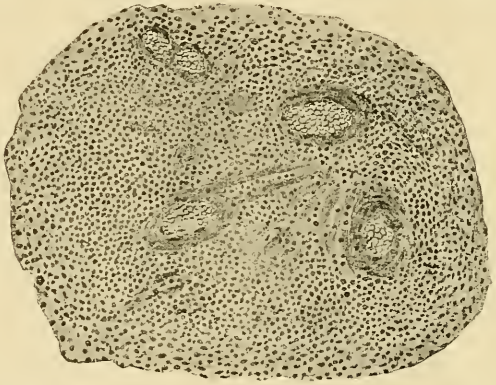


FIG. 66.—One of the foci (F of Fig. 65) more highly magnified (after Oppenheim).

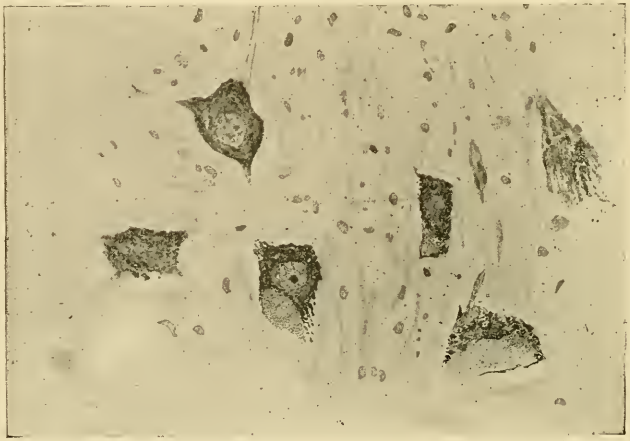


FIG. 67.—Swollen and disintegrating cells in a case of myelitis

softened tissue also contains axis cylinders in various states of disintegration.

In many cases of diffluent myelitis these changes of the individual elements of the cord are the only ones that can be distinctly made out. But in the parts directly surrounding the focus of most intense inflammation further changes can be made out after proper hardening and staining with the various dyes. On such sections the blood-vessels will be found dilated, and innumerable leucocytes can be seen in the vicinity of such vessels. The nuclei of the smaller arteries and capillaries will be found to be enormously increased and in a state of proliferation. The sheath of the blood-vessels is very much distended, blocked in part by the coagulation of blood, and round blood-corpuscles may even be found in the adjacent tissue.

In the gray substance the large nerve-cells are swollen and granular, many of these granules showing distinct evidences of degeneration. The processes of the cells are either shrivelled up or entirely lost, the contour of the cells less distinctly defined, and changed from the polygonal form to spherical or oval-shaped bodies. The neuroglia of the gray matter will appear denser than under normal conditions. In the white substance similar changes will be found in the blood-vessels and in the interstitial tissue. The white substance often has a distinctly fibrous appearance, containing many spider-cells or cells of Deiters. The white nerve-fibres themselves undergo degeneration. The axis cylinders are irregular, swollen, and often transversely divided. In some cases the entire nerve-fibre is destroyed or disintegrated and the space once occupied by such fibres is left vacant or occupied by granular matter. In some cases of myelitis the nerve elements are more intensely affected than the interstitial tissue, while in others the changes in the interstitial tissue are the more prominent feature in the cross-section, and the nerve-tissue has evidently been destroyed secondarily. In transverse myelitis the changes may be distributed equally through the entire cross-section; in other cases the changes may be more intense in the gray than in the white matter, and in some more intense in the ventral half than in the dorsal half. In cases of meningo-myelitis, particularly in those of traumatic or specific origin, the most marked changes are near the periphery. Here the pia will appear thickened and the morbid changes can be traced along the connective tissue passing from the pia into various portions of the cord. If the myelitis is of the disseminated order, small foci of disease may appear in various portions of the cross-section and in various segments of the cord, intervening parts maintaining a tolerably normal appearance. If the myelitic changes are most prominent in the vicinity of the central canal and the parts surrounding it, we speak of a central myelitis; but it is rare to find such central myelitis without some additional symptoms of a diffuse inflammatory process. The nerve-roots in connection with the inflamed segments are, as a rule, altered, and will present appearances somewhat similar to those found in the white matter of the cord. The vessels are dilated, the nuclei and the tissues about these vessels exhibit various degrees of proliferation, the myelin

is disintegrated, and the axis cylinder either swollen or distorted; but these degenerative changes can, as a rule, be traced only a short distance from the diseased cord.

Secondary changes follow upon the area of inflammation, and the tracts will be affected in an upward or downward direction, according to the direction in which they transmit impulses. Thus, after a transverse myelitis the lateral columns will degenerate downward throughout their entire extent, but an ascending degeneration will occur in the parts that transmit impulses in a centripetal direction. (Fig. 64.) Among those exhibiting ascending degeneration are the posterior columns, the cerebellar tract, and the antero-lateral ascending tract. The inflammation spreads a short distance upward and downward by contiguity, and those parts which one would suppose to be subject to descending degeneration only may be affected for a short distance above the lesion; but such changes are of a distinctly inflammatory character and altogether different from the purely secondary changes, which rarely offend against physiological principles. These degenerations are, as a rule, developed very promptly after a transverse lesion, and often continue to exist after the initial inflammation has pretty well disappeared.

Whether fibres that have once been seriously altered, or even destroyed, can ever regain their function or can grow anew, is a matter of serious doubt, and yet recovery takes place in a fair number of cases in which absolute paralysis, with signs due to descending degeneration, had existed for a number of months, or sometimes for a year or more. We must suppose in such cases that some fibres were so little altered that when the inflammatory products were absorbed they still retained the power of conduction, and it is more probable that such fibres may be restored to absolute health than that entirely new fibres can be formed within a nerve-sheath, or that destroyed fibres can be replaced by new ones.

Differential Diagnosis.—The more or less acute onset, the often sudden loss of power, the rapid spread of anæsthesia, the permanency of all these symptoms, together with the retention of urine and fæces, and the flaccid and atrophic symptoms at the level of the injured part, together with the spastic symptoms in the parts supplied from segments below the level of the lesion—all these symptoms will leave little doubt of the diagnosis of acute myelitis. In addition to this the etiological factors in the case—the occurrence of traumatism, a preceding syphilitic infection or pre-

ceding bone disease, or marked infectious influences—will help to corroborate the diagnosis.

Acute myelitis may resemble hemorrhage of the cord, but in cases of hemorrhage the onset is more sudden than in cases of acute inflammation, all the symptoms being developed within a very few minutes. There is, as a rule, too, much more pain than in cases of myelitis; but hemorrhage is frequently enough the first stage of a myelitis, and if symptoms indicating a spread of disease follow upon what is supposed to be an initial spinal hemorrhage, it is fair to conclude that a myelitis hæmorrhagica has followed upon the initial extravasation of blood. Direct injury to the spinal column may be another factor tending to corroborate the diagnosis of hemorrhage.

A rapidly ascending myelitis may suggest the acute ascending (Landry's) paralysis, but in cases of myelitis the progress will be clearly from the level of the first injury, and is not apt to attack the parts in succession from below upward, including the trunk, as in cases of Landry's paralysis. In ascending myelitis, moreover, sensation is disturbed from the start, and all the trophic, as well atrophic, symptoms are much more characteristic of a myelitis than they are of Landry's paralysis. But if the myelitis begins in the lumbar portion of the cord, and gradually spreads upward, the difficulties of diagnosis may be extremely great. Landry's paralysis is unusually rare in children, whereas myelitis is relatively frequent.

The distinction between meningitis and myelitis is not of great practical importance, for meningitis is rarely present without some involvement of the cord, and if the symptoms are purely meningeal, they are generally associated with other symptoms pointing to a wide-spread affection. A primary spinal meningitis is a great rarity, except as a part of cerebro-spinal disease or after disease or injury of the spinal column. The involvement of the meninges in a given case will be indicated by considerable pain in loco morbi, and by the presence of distinct neuralgic pain along the nerves emanating from the diseased portion of the cord.

The question at times arises whether a case is one of myelitis or multiple neuritis. In the latter symmetrically located pains are a more prominent symptom, trophic disturbances are not so marked as in myelitis, and the symptoms never include vesical and rectal disturbances. Moreover, we never have that combination of paralytic and spastic symptoms which we so frequently find in cases of myelitis. In cases of multiple neuritis affecting all four extremities the symptoms in all the extremities are entirely the same, whereas in cases of myelitis they would be of the flaccid order, say, in the upper extremity, and of spastic order in the lower extremity.

A more difficult task it is to distinguish between a myelitis and hysterical paralysis, and yet a careful examination of the patient should reveal important points of diagnosis. Thus, in hysterical paralysis, legs that cannot be used in standing or walking may be moved freely in bed; the rigidity is not so marked as in cases of myelitis, and if present can be more easily overcome, as a rule, than in myelitis. Bed-sores are rarely present in hysteria, and anæsthesia, if present, is anomalous in distribution. The reflexes, too, are not so distinctly exaggerated in hysteria as they are in cases of myelitis. If the symptoms should point to a lumbar affection, the lack of atrophy and the persistence of the knee-jerks will help to differentiate the hysterical paralysis from a spinal paraplegia. The bladder and rectal symptoms are also not so marked in hysterical as in myelitic cases. The very suddenness of the onset in hysterical cases, the fact that the paraplegia is frequently due to a sudden fright, or a deep emotional condition, may also point to hysteria rather than to myelitis. But in all such cases the fact that hysterical subjects may suffer from organic lesion should be borne in mind.

Prognosis.—The prospects in cases of acute myelitis will vary according to the level affected. Cervical myelitis is naturally a more serious disease than myelitis of lower portions of the cord, for in the former an extension upward to the respiratory and cardiac centres constitutes one of the grave possibilities of the case. In these, as well as in dorsal and lumbar myelitis, the danger to life arises chiefly from the complicating conditions, particularly from bed-sores, from cystitis, and pyelo-nephritis. The earlier these symptoms set in the graver the prospect of the case; but not a few of such cases get well in spite of all complications, and if the myelitis can be proved to be due to specific disease, or to some other form of mild toxic infection, recovery is more probable than in the traumatic cases of myelitis or those in which the etiological factor was entirely unknown.

The prognosis will also vary according to the intensity of the affection and the extent of cord involved. If all the symptoms are developed rapidly, then become stationary

and show not the slightest sign of improvement for weeks or months, the probability of spontaneous recovery is extremely slight; but any improvement which sets in, either in the form of diminution of anæsthesia, of the disappearance of bed-sores, or of a slight gain in motion, is a hopeful sign of greater improvement later on. I have myself seen complete recovery in cases of myelitis in which the palsy was absolute for a period of nearly six months, with marked contractures and increase of the reflexes and with slight vesical symptoms, but I cannot recall a single case in which complete recovery set in if deep bed-sores developed at an early day, and marked cystitis appeared very early in the disease, the only exception to this rule being in cases of distinct specific myelitis.

TREATMENT.—In the treatment of myelitis the following plan should be pursued if the patient is seen during the acute stage. An ice-bag should be applied to the greater part of the spinal column; counter-irritation may be used, but the danger of trophic changes in the skin should be remembered, and such trophic changes should not be encouraged or started up by an excessive use of counter-irritants. The patient should be placed absolutely at rest, if possible with some form of extension. His bowels should be thoroughly purged, best by the use of calomel, and the bladder catheterized by careful hands and watched for the first signs of a cystitis. (If cystitis should develop, no time should be lost, even during the acute stage, in beginning the usual treatment for such conditions.) All these measures will tend, first, to make the patient more comfortable, and, secondly, they will surely lessen the danger from complicating conditions. The diet should be of a mild, non-irritating kind, and the kidneys should be encouraged to greater activity. Under such conditions the administration of small doses of digitalis or of the acetate of potash will be quite in order. Ergot was recommended years ago by Brown-Séquard, and may be administered with the idea of limiting the area of inflammation. I cannot say that I have ever seen any direct results from ergot, but it seems to do no harm, and considering the seriousness of the disease it may well be tried. In cases in which there is reason

to think that there is much inflammatory exudation, and particularly in those of specific origin, the administration of the iodides is quite in order, or of the mercurials and iodides combined; but if mercurials are exhibited it is useless to give them in any other way than by inunction.

The main objects are to prevent the serious complications so common in these diseases, and to give the diseased organs a fair chance of spontaneous recovery. It is of the utmost importance, therefore, in cases of myelitis, to keep the patient absolutely clean—a task not so easy in view of the frequent dribbling of urine and of the involuntary passage of fæces. The child should be placed upon a water-bed. Nurses should, invariably, receive instructions to keep the bed-linen absolutely smooth, and to promote this end it will be best to have the bed dusted very liberally with some slightly aseptic powder. In the case of female patients, pads should be put in place to catch the dribbling urine and to prevent its soaking adjacent parts; in the case of boys, urinals should be used from the start. It is a common practice with me to order the patient's position in bed to be changed at least every hour, so that no one part is pressed upon for too great a length of time. If in spite of all these precautions bed-sores should form, these should be treated according to the best surgical principles. Latterly I have been in the habit of dusting the sores with dermatol or aristol and covering them completely with light antiseptic dressing, shielding the dressing as well as may be from contamination by urine or fæces. It is better to change the dressing frequently than to allow any infection of the sores through uncleanliness. In hospitals or among the poorer classes less expensive substances, such as bismuth, may be used; and if cystitis has been set up, the bladder should be washed several times a week with some weak antiseptic solution.

After the symptoms of the acute stage have been successfully treated, and the patient has passed into a more or less chronic condition, the question arises as to the proper treatment of this latter stage of the disease. I am in favor of making the attempt again and again of affecting the focus of inflammation by the administration of the

iodides. In nine cases out of ten this will be unavailing, and yet there is no good reason why the effort should not be made. But the possibility of recovery should be well weighed in the scales as compared with the gastric disturbance which these drugs so often excite. The nutrition of the child must be maintained at all odds, and it is far better to abandon the iodides than to permanently impair the assimilation of food. If the iodides cannot be given, inunctions of the oleate of mercury or of the usual mercurial ointment should be substituted for the iodides. Counter-irritation may be attempted, either by the cautery, by blistering, or by mere cupping; but little direct good is to be expected from these remedies. The child should be kept absolutely at rest, and the spinal column should be disturbed as little as possible. Electricity applied to the spinal cord is of very doubtful utility, but there is all the more reason for using it in the treatment of the paralyzed parts. It does excellent service here as a form of exercise for the maimed limbs. Massage has very much the same, and, I believe, a better, effect than electricity. It tends, in addition, to maintain the nutrition of the parts, and to overcome the tendency to contractures, which so frequently give rise to the most disagreeable symptoms in these cases; but neither electricity nor massage should be pushed if the involuntary spasmodic contractions become more frequent, as they often do in consequence of these measures. These contractions are not harmful, but are extremely irritating to the patient, and if inordinately increased may disturb his rest, and thus interfere with the general nutrition.

As soon as the child has sufficiently recovered, it is of the greatest importance to give it all the fresh air possible, and, if necessary, to provide it with a wheeled chair, so that it can be given its regular outing. Its diet must be carefully looked to, and all unnecessary excitement should be avoided. Tonic measures may be employed; and if the child is in the charge of a competent person give lukewarm baths, followed by cool or cold douches of the spine, and let this be done before the massage is given. Iron, quinine, or arsenic may be administered for their general tonic effect. Strychnine may be given in very small doses in those

cases in which there is reason to think that the substance of the cord has not been absolutely destroyed, and that the function of the diseased parts could be increased by the use of this drug. I know that it is an extremely popular drug in these diseases with all physicians who are not specialists, but I have found it to be a double-edged weapon. If it increases nerve conduction for a time, such improvement is very apt to be followed by a further diminution of function, and in other cases again it produces annoying muscular contractions, which are as disagreeable as those that result from an excessively strong electrical current. In these cases, too, the physician who watches his patient carefully, and does not attempt to do too much, will succeed far better than he who is continually meddlesome and ever anxious to change treatment.

INJURIES OF THE SPINAL CORD.

The direct results of jumping, of falls, and of injury due to falling weights are, on account of the more delicate nature of the spinal vertebræ, more serious in childhood than in later years. A fall downstairs, or a simple fall out of bed, or a mild blow inflicted by another child, may be a sufficient cause to start the symptoms pointing to actual injury of the spinal cord.

The symptoms of spinal-cord injury may be extremely varied. I do not propose now to discuss those cases in which traumatism is the remote and questionable cause of a subacute or chronic form of spinal disease developing months, or even years, after an accident; but I wish particularly to direct attention to those cases in which the spinal injury is followed immediately, or within a period of a few weeks, by symptoms which point to some traumatism. In the severest form of injury to the cord there is immediate paralysis both of the motor and sensory functions. Inasmuch as the injury more frequently involves the dorsal and lumbar portions, a spastic paraplegia with anæsthesia of the lower extremities, with loss of vesical and rectal control, constitute the chief symptoms which may be developed within a few minutes or within a few hours after the injury.

If examined more in detail we can determine by the character of the symptoms, first, the exact portion of the cord involved by the injury, and secondly, the amount of injury done at any level. The question of the exact extent of the injury up and down the cord is determined by the parts paralyzed, and more particularly by the extent of the anæsthesia. These cases are indeed well calculated to illustrate the principles of spinal localization. In cases of cervical lesion the upper as well as the lower extremities are involved. In the upper extremities the paralysis is of an atrophic order, while spastic paralysis is present in the lower extremities. The anæsthesia in-

volves both the upper and lower extremities as well as the trunk, to or from a level supplied by the nerves coming off from the injured segment. In cases of cervical injury the sympathetic may be involved. In cases of injury to the dorsal region, which are the most frequent, the arms are not affected, the lower extremities are in a condition of spastic paralysis, the bladder and rectum may be involved, and the anæsthesia extends from below upward to a level corresponding to the segment or segments injured. The band of hyperæsthesia, or the upper level of the anæsthesia, will correspond to the upper limit of injury. If the injury has been done to the lumbar enlargement, the paralysis of the legs is of a flaccid order, the reflexes are diminished or lost, the vesical and rectal reflexes lost, and the areas of anæsthesia will vary according to the segments involved. Injuries to the cauda equina are of particular interest because of the peculiar character of the anæsthesia, upon which alone an accurate diagnosis can be based.*

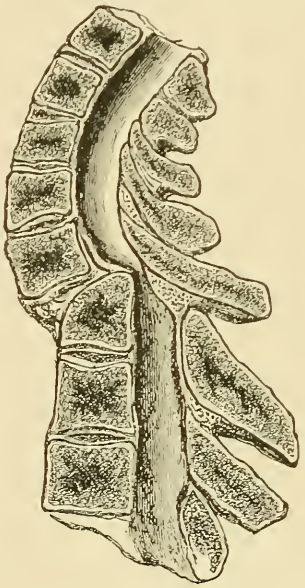


FIG. 68.—Sketch of Section of Spine in a Case of Fracture Dislocation of the Seventh Cervical Vertebra. (After Thorburn.)

The exact study of the symptoms will often help us to determine the upper as well as the lower limits of injury. Thus in one case which I have had occasion to observe, there was spastic paralysis with slight diminution of the knee-jerks, but presence of ankle clonus. There was spasmodic twitching of the legs, loss of vesical and rectal reflexes, as well as a tendency to bed-sores. The difference in the behavior between the knee-jerks and the ankle clonus proved conclusively that the injury had slightly involved the upper lumbar segments, chiefly the lower dorsal region, but that every part of the cord below the upper lumbar segment had escaped injury, or else the ankle clonus would surely not have been present. In determining the extent of the cross-section involved, a very important question, and one which helps us to decide whether a complete crush has taken place or not, we must keep the physiology of the cord in mind, and endeavor to make out whether the anterior horns as well as the lateral or the posterior columns have been involved. Thus, in a case in which the lesion was in the lower dorsal region, and in which there was complete anæsthesia with marked spastic paralysis, I found that the muscles of the back showed, after a lapse of weeks, neither a tendency to atrophy nor to changes in electrical reactions. I argued from this, and correctly too, as the event proved, that the chief injury was done to the posterior and lateral portions of the cord, and that the ventral portions had entirely escaped.

* See the paper by Starr in American Journal of the Medical Sciences, July, 1892.

Another aid to accurate diagnosis of the lesion will be found in the sensitiveness to pressure over the spinal column at the seat of injury. The parts that are sensitive should correspond to the segment of the cord which an examination of the patient has shown to be diseased, and before coming to any definite conclusion the physician should remember the relation of the external parts to the segments of the cord. If these two sets of facts do not accurately correspond, the preference should, to my mind, be given to the seat of injury as determined by the study of the paralysis and the anæsthesia; but if the level determined in this way is not far distant from the seat of pain on pressure, both should be included within the area to be operated upon.

The course of the disease will depend largely upon the region affected and upon the extent of injury done. Injuries to the cervical region are, on the whole, more serious than those to the dorsal and lumbar portions of the spinal cord. Injury to the lumbar spine is generally followed by more serious symptoms than is the case after dorsal injury. If the initial symptoms indicate a comparatively slight lesion at any level, the progress of the disease is apt to be more favorable than if a complete or nearly complete crush of the cord has occurred. If the symptoms show a tendency to improvement after a few weeks, or after a month or more, the possibility of complete recovery may be considered; but if they remain stationary for a long period without the slightest indication of improvement, actual recovery is rare, unless relieved by operation.

The danger to life is greater in the cervical cases, in which the proximity to the vital parts is of much importance; and in severe cases of dorsal or lumbar injury the complicating conditions, such as cystitis and bed-sores, may bring about a rapidly fatal issue. After an initial injury that is relatively slight, degeneration may set in, which will be characterized by the onset of rigidity and contractures, and from the onset of these symptoms the prognosis as regards complete recovery may become very much graver.

PATHOLOGY.—The actual anatomical changes in traumatic lesions of the spinal cord may vary greatly. Even without injury to the vertebral column hemorrhage may occur from the effect of the shock, and this may be either epidural or subdural. The probability of the seat of the hemorrhage will have to be argued from the general character of the symptoms. In persons whose arteries are fragile, hemorrhage is much more likely than in persons whose vascular system is entirely normal. Persons with syphilitic disease will, therefore, be much more liable to traumatic hemorrhage than those not so affected. If the hemorrhage is considerable, whatever its location may be, the cord will suffer from compression and may undergo softening unless the blood that is exuded is rapidly absorbed. In all cases, inflammatory products, which may be tinged by blood, will be found at the seat of injury. At times, in spite of a sudden paralysis developed immediately after an injury, no tangible lesion can be discovered at the time of operation or on the post-mortem table. We must then suppose either that the evidences of the initial lesion have disappeared, or that the traumatism has resulted in functional changes, or in such, at least, as are beyond the discovery by our present

methods. If examined months or years after the initial injury, the cord may present nothing but the ordinary symptoms of chronic myelitis with considerable shrinkage and wasting of the entire substance of the cord. Such wasting may be at times more marked in the white columns, at other times more distinct in the gray matter. In addition to the local changes found, the cord, if examined carefully, will reveal ascending and descending degenerations in accordance with the intensity of the process at any given level. I have seen several cases of spinal injury with severe spinal symptoms in which at the time of the operation no tangible changes were found in the cord, but the cord was evidently compressed by inflammatory exudations that had collected between the bone and the dura. Adhesions also form under these conditions between the dura and the surrounding parts. The impairment of function is, therefore, due to extra-spinal conditions. The breaking up of such adhesions and the removal of such inflammatory products are followed by improvement in the condition of the patient, if such removal is effected within a relatively short period of time after the accident.

TREATMENT.—In cases of serious injury to the spinal cord, absolute rest is essential. The case may be treated in every respect for the first few days as though it were a case of non-traumatic myelitis, that is, by application of cold to the spine and by extension. As soon as the condition of the patient will permit, a careful examination should be made in order to determine the amount of injury done. The details of the treatment will not vary from that advised in cases of myelitis. The old habit of using the actual cautery, of blistering, and the like, is not to be recommended, for little good can follow it. As was said in the case of myelitis, the danger of trophic disturbances in the skin is great enough without such additional encouragement. The most important question that arises is whether anything can be done for the patient by surgical means. There is still much hesitation on the part of physicians and surgeons in this respect.

If the patient is in good general condition, and is able to stand the shock of the operation, the advisability of such surgical interference should be considered purely upon the merits of the case, and if the evidence points to the fact that injured bone is pressing upon the cord, or that severe hemorrhage has occurred and that there is danger of permanent harm to the cord, the surgeon should be permitted to expose the injured region. This can be done with considerable impunity by competent surgeons at the present day, and, as I have said, should be done as early as practicable after the injury. The cases in which surgical interference is useless are those in which all the symptoms indicate absolute crush of the cord, or in which the rapid development of all the symptoms points to a probably fatal issue; or those in which so long a period of time has elapsed since the injury that there is no good reason to believe that the conditions can be relieved by operation. I have advised operations upon the spinal column in four cases, two of which were in children under the age of fifteen. The results were satisfactory, although neither one attained complete recovery; but I attribute this to the fact that the cases were sent to me at too late a period after the accident. The operations are not nearly so dangerous as those upon the brain, and

much encouragement may be derived from the statistics of various surgeons who have not lost a single case operated upon for serious spinal injuries. If for some reason or other the operation has not been performed and the patient is left with a chronic spinal disease, the attempt may be made to benefit the patient by the administration of the iodides; and the crippled condition of the extremities may be somewhat improved by ordinary surgical and orthopedic measures, such as have been frequently referred to in the discussion of myelitis and other forms of palsy.

CHAPTER XVI.

SYPHILIS OF THE SPINAL CORD: SPECIFIC MYELITIS AND MENINGO-MYELITIS.

In the adult, syphilis of the spinal cord can now be recognized by a definite combination of symptoms. This is due to the work of Erb, Gowers, Marie, Oppenheim, Siemerling, Hoffmann, the author, Nonne, and others. In the child syphilitic disease of the spinal cord is not nearly so frequent as in the adult, but I do not hesitate to devote a special section to this subject, both because I think such cases can be easily overlooked and because they present a number of points in differential diagnosis which are of the greatest interest and which must be carefully considered if grave errors in diagnosis are to be avoided.

Symptoms.—In former days it was customary to make the diagnosis of syphilis of the spinal cord if the symptoms pointing to spinal lesion were irregular and would not fit into any of the ordinary types of spinal disease. As a matter of fact the nature of the morbid process underlying these syphilitic spinal diseases is such that irregularities in distribution, and in the development of the symptoms, are very apt to occur, and we should be able to recognize the symptoms of syphilitic disease whether or not the patient reveals, or we can prove, previous syphilitic infection.

In all but a few of the cases the onset of the disease is gradual. By degrees the legs or the arms, or both the lower and the upper extremities, which have shown some weakness, become paralyzed. One leg, or one arm, is at times more paralyzed than the other. This paralysis may be of the atrophic kind, but is much more apt to be of a spastic order. It is often associated with intense pain (par-

aplegia dolorosa), or with anæsthesia—the anæsthesia and paralysis may be crossed (Brown-Séquard type). The reflexes are generally increased, rarely absent. If the specific process is situated in the cervical segments there are atrophic paralysis with loss of reflexes in the upper extremities, spastic paralysis with rigidities and contractures in the lower extremities. The vesical and rectal reflexes may be interfered with. If the dorsal or lumbar segments are involved, the symptoms will closely resemble those following upon myelitis of the respective segments; trophic disturbances may occur; bed-sores may develop; in short, we may have all the symptoms of a wide-spread spinal affection. It is evident that if we wish to distinguish between specific disease of the cord and the various forms of acute or chronic myelitis we must look for some distinct points of differential diagnosis.

Erb, referring to be sure to the conditions in the adult, has established a type of spinal-cord disease which he proposed to call syphilitic spinal paralysis. This special type bears the following characteristics: First, the usual symptoms of spastic paraplegia, with its peculiar gait, carriage, and movements; second, marked exaggeration of the deep reflexes; third, muscular contractures, which are slight as compared with the exaggeration of the reflexes; fourth, involvement of the bladder; fifth, a slight yet distinct disturbance of sensation; sixth, gradual onset of the disease; seventh, a decided tendency to improvement. There is no doubt that this type of spinal disease, so well characterized by Erb, does occur. The same series of symptoms has been recognized by Rumpf, and since Erb's publication cases of this description occurring in children have been observed by Friedmann and by myself. But I have taken some pains to prove that there are other types, quite as frequent as this one, and that it is a great mistake to hesitate in making the diagnosis of spinal-cord syphilis unless the symptoms of Erb's type are present. It is the author's conviction that, if we wish to make a positive diagnosis of syphilis of the spinal cord, we should pay attention to the following points, and not exclusively to those presented by Erb as characteristic of the special type he has described.

First, the most striking feature of syphilis of the spinal cord is the unusual distribution of the disease over the greater portion of the cord, involving, as it often does, the cervical and dorsal, as well as the lumbar enlargements. Second, the slight *intensity* of the morbid process at one level as compared with the *extensive* area involved, as evidenced by the preservation of some of the functions of the cord with complete loss of others. Third, the rapid dwindling of some of the symptoms and the very chronic persistence of others. Thus in some of my cases the anæsthesia lasted but a very short time, while the paralysis was recovered from with extreme slowness. Fourth, the very frequent history of other symptoms pointing to specific disease in the same or distant parts of the central nervous system.

To emphasize these views let me state that I should be inclined to suspect specific disease of the cord if the patient presents symptoms of paralysis, whether they be of the spastic or flaccid character, and whether the contractures be slight or not, provided he furnish evidence of a morbid process affecting a very large part of the cord, and yet showing a relatively slight intensity at any given level of the cord. He may, for instance, exhibit the symptoms of extreme paralysis, spastic or atrophic, with partial or slight anæsthesia of the parts paralyzed, with little or no involvement of the bladder; or, as often happens, he may present traces of specific disease in other parts of the central nervous system. The chief difference, according to this, between the ordinary forms of myelitis and the specific diseases of the cord can be understood if we remember that the symptoms of an acute or subacute myelitis prove that the entire cross-section of the cord is affected almost simultaneously and to an equal degree, whence it follows that in such cases severe paralysis is likely to be associated with severe anæsthesia, with marked contractures, with absolute loss of vesical and rectal control, with serious trophic disturbances, and so on; whereas, in the cases of spinal syphilis the morbid process invades the cross-section of the spinal cord partially and slowly. We may, therefore, find symptoms which point to a very marked affection of one or more of the systems of the spinal cord and to relative immunity from disease of the gray matter or other portions of the cord. Thus we may have extreme paralysis, but only slight anæsthesia; or extreme loss of power with relatively slight rigidity, as Erb pointed out. Furthermore, in the ordinary cases of myelitis, the symptoms point to a certain portion of the spinal cord at which the disease is most intense, while in cases of syphilis of the spinal cord the clinical symptoms show that the disease involves a very large portion, if not the entire spinal cord, yet affects each single segment but relatively little. A still further aid to differential diagnosis is the very frequent involvement of the brain, at the same time that the majority of the symptoms point to disease of

the spinal cord, or if these two sets of symptoms do not set in simultaneously, we frequently have in a patient who presents symptoms of a specific myelitis the history of a preceding illness in which the symptoms were of a cerebral rather than of a spinal character. (Unequal ocular, and, above all, unequal pupillary symptoms, are very common symptoms of cerebral syphilis.) The rapid and often unexpected recovery, as well as the relapses, help also to distinguish these cases from the usual forms of myelitis.

In view of the rarity of these diseases, or possibly of the failure to recognize them when they do occur, I wish to give a few typical cases. The first one I take from Friedmann's article on relapsing, probably specific spastic, spinal paralysis in childhood.

A boy, five years of age at the time of examination. Nine months before the birth of this child the mother miscarried with twins at the end of the second month of pregnancy. The birth of the boy was entirely normal, but the head was said to have been very large. Relative reduction of the size of the head in the next few months. Four weeks after birth a skin eruption appeared which covered the entire body; it was vesicular at first; later on it ulcerated, and terminated in desquamation. The child began to exhibit normal mental development, learned to talk at the end of one year, and began to teeth at the age of six months. At the age of three months there was distinct difficulty in moving the arms and legs. Three months later the right arm could be moved, and at the age of one and a quarter years all four extremities appeared to be entirely normal. In the second year, the child having learned to walk well, paralysis again gradually developed in the *left* arm, and disappeared once more after six weeks. From the second to the fourth year the boy was healthy, and passed through measles and diphtheria without any disagreeable sequelæ. In the fourth year he complained much of headache, particularly in the occipital region, and gradually his gait became weaker and weaker, and he began to drag the right leg, frequently falling in the attempt to walk. There was slight difficulty in micturition and the legs were somewhat rigid. In this attack the arms were entirely free, and there was no history of spasms or convulsions at any time. The patient was in this condition when examined by Friedmann. The only other points of interest in the case were the very bad condition of the teeth, slight increase of the reflexes, and entirely normal sensation.

More recently J. Hoffmann has reported the case of a boy, who, at the age of twelve years, developed a typical spastic paraplegia, evidently due to hereditary syphilis, manifest signs of which appeared in the first years of life.

The following case, observed by myself, presents many similar features:

E. S——, a girl, aged six years, born in this country, of German parents, was brought to my clinic in June, 1893. The mother gives a history of protracted labor, but child was entirely normal; began to cut teeth at six months, to stand and walk at the age of one year, and learned to talk well before she was two years old.

The child had been perfectly well, with exception of mild attacks of whooping-cough and measles. At the age of five years the mother noticed that the child began to walk in a peculiarly stiff manner, and that its mental development was somewhat retarded. On examination we noted spastic paralytic gait; spastic paraplegia of lower extremities, more marked on left side; left upper extremity slightly paretic and rigid; both knee-jerks exaggerated; triceps and wrist reflexes lively on left side. Pupils unequal; left pupil dilated and does not react to light; right pupil reacts sluggishly to light; both pupils react sluggishly during accommodation. No sensory symptoms.

The suspicion of syphilitic disease was strengthened by an examination of the mother. She has had two miscarriages; three children died in early life. Five years ago (at the age of thirty years) she had left hemiplegia; no loss of consciousness; recovered in a few weeks; has distinct "rheumatic" attacks; gets dizzy while washing her face in the morning; her pupils are unequal; no reaction to light or during accommodation; knee-jerks absent; slight Romberg symptom; no bladder trouble; delayed sensory perception in the lower extremities.

Thus we have a tabic, if not a purely syphilitic affection in the mother, and spinal syphilis in the child.

Differential Diagnosis.—It seemed to me best to introduce most of the salient points of differential diagnosis in the description of the symptoms of the disease, and the comparison with myelitis was inevitable. On the distinction between myelitis of the ordinary types and specific disease of the spinal cord, I need say nothing more. Syphilis of the spinal cord might be confounded with a spastic infantile palsy, particularly with a spastic diplegia and paraplegia resulting from meningeal hemorrhage occurring during the period of labor; but in these latter cases the trouble can be traced distinctly to the earliest period of life, and there is never any history of relapses, and rarely of any marked improvement followed by relapse. Moreover, in the typical cerebral palsies the contractures are apt to be more extreme, and defective cerebral development is much more frequent than in the syphilitic cases coming on subsequently to the birth of the child. An acute infantile cerebral palsy might possibly simulate specific disease,

were it not for the far greater frequency of the hemiplegic form of paralysis in these cases than in the purely syphilitic types of disease. Special difficulties might, however, arise in cases in which the acquired infantile hemiplegia would be proved to be due to syphilitic disease of the arteries, and under such conditions a combination of spinal syphilitic disease with cerebral disease would not be impossible, although I have not yet come across a case of this description. Spinal syphilis in a child should not be con-

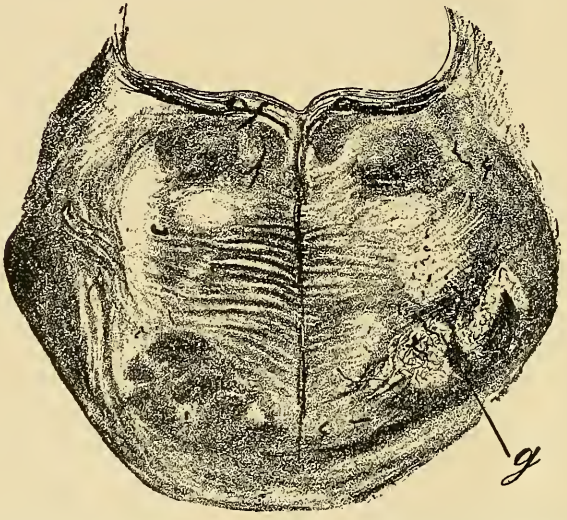


FIG. 69.—Case of Multiple Cerebro-spinal Syphilis. Section through pons showing gummatous formation (*g*) in ventral portion, with considerable destruction of tissue. Weigert's hæmotoxylin stain ; low power.*

founded with hereditary spastic paralysis; possibly the latter may be developed on a specific basis.

MORBID ANATOMY. — The anatomical changes in the spinal cord, due to syphilis, may be quite as varied in the child as in the adult. It is well known that syphilis is apt to cause disease of the blood-vessels. Obliterative endoarteritis, with subsequent softening of the area supplied by the diseased vessel, is perhaps the best known anatomical process directly attributable to syphilis; but a general ar-

* Figs. 69–71 are reproduced by the courtesy of the editor of the New York Medical Journal.

teritis is quite as frequent as an inflammation of the endothelium alone. Bruce has directed attention to changes in the adventitia (nodose periarteritis); moreover, veins are subject to syphilitic changes quite as often as the arteries are (phlebitis obliterans, Greiff). All these vascular changes are much more pronounced in the pial covering than in the substance of the spinal cord.

The investigations of other authors, as well as my own,

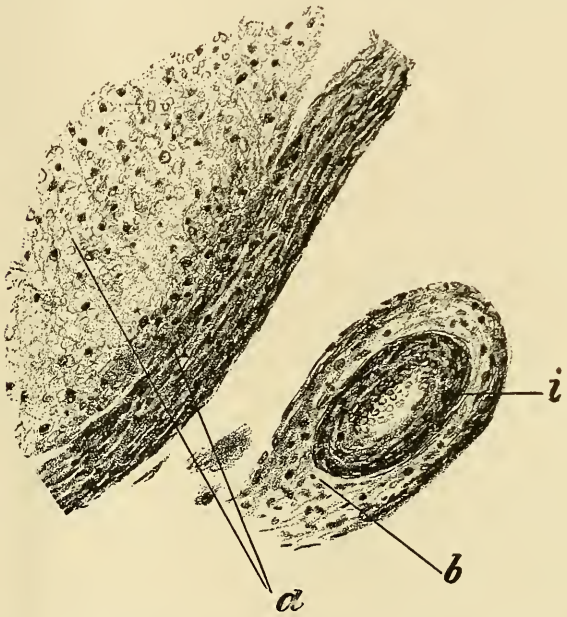


FIG. 70.—Section from Ventral Surface of Medulla Oblongata (high power), showing Infiltration of Pia and Substance of Medulla (*a*) and Typical Syphilitic Arteritis; Marked Thickening of, and cellular proliferation in Intima (*i*); Narrowing of the Lumen; Cellular Infiltration of Adventitia (*b*).

have shown that syphilis of the spinal cord is more often associated with a subacute or chronic meningitis, or meningo-myelitis, than with any other process. The disease starts, as a rule, in the pia, and subsequently invades the spinal cord. There can be little doubt, however, that the changes may in some instances be developed in the reverse order.

In the gross specimen we find the pia thickened and

often covered by a thick gelatinous substance. On microscopical examination this thickening is seen to be due to a proliferation of all the tissues. The cells are multiplied in number, the nuclei have increased, and the blood-vessels show the characteristic changes of specific arteritis. The walls of these vessels are thickened, all of the coats participating to an equal degree. The process very often starts in the intima, but not invariably so, as was maintained some years ago by Heubner. The pia is generally adherent to

FIG. 71.—Section through a Portion of Dorsal Cord (Ventral Surface). Marked thickening of pia; cellular infiltration of same, seen best in that portion which projects inward; infiltration of substance of cord.

the spinal cord, and the cellular infiltration extends from it into the substance of the spinal cord. This invasion is a very gradual one, and may begin at almost any point of the cross-section of the cord; but the lateral columns are more frequently involved than any other region, and for this reason the spastic symptoms are, as a rule, the first to be developed. In other cases in which the morbid process invades the posterior columns the symptoms may resemble those of tabes rather than those of a spastic form of paral-

ysis. If the invasion occurs from the ventral surface, as it rarely does, the symptoms will naturally be of an atrophic order rather than of a spastic or sensory kind. But wherever the first point of attack may be, the morbid process gradually works its way inward from the periphery, and its plan of attack is generally from symmetrically situated points. The cellular proliferation and the slight increase in neuroglia tissue, as well as the gradual disappearance of the integral elements of the cord, give to these cross-sections, if examined under the microscope, the appearance of ordinary myelitis, and if it were not for the characteristic changes in the blood-vessels and for the evidence that the process has worked its way inward from the periphery, and, indeed, from the pia, the anatomical proof of a specific process would be very difficult to establish. As it is, there are not a few cases in which satisfactory proof cannot be given, and the pathologist is compelled to rely upon the clinical evidence in the case to prove the syphilitic nature of the disease. But the wide-spread character of the disease, and the very gradual destruction of the different systems* of the cord, as well as the important part played by the pia and the blood-vessels, leave little doubt as regards the true nature of the inflammatory process, and at the same time help us to understand the peculiar behavior of the clinical symptoms. The Figures 69–71 will give full details of the specific meningo-myelitis as seen in one of my adult cases.

The meningo-myelitis may be associated with special gummatous deposits in any part of the cord or in the brain. Thus, in one of my cases a gumma in the pons was associated with a wide-spread specific meningo-myelitis at the base of the brain and throughout the entire extent of the cord. If such a complication exists the child may present the symptoms of tumor of the brain or cord, together with the symptoms of extensive meningo-myelitis.

The fact that spinal syphilis has a distinct tendency to improvement makes the prognosis favorable, although this tendency implies also a danger of relapses. But syphilis in

* Nonne has shown that Erb's type may be due to a combined systemic degeneration involving the pyramidal, the cerebellar tracts, and the tracts of Goll.

the spinal cord of the adult, as well as of the child, is amenable to treatment, and the prognosis is distinctly more favorable than in cases in which the same symptoms might be present and not due to syphilis. The prognosis is, for instance, very much more favorable than in cases of congenital diplegias or paraplegias. The possibility of recovery should therefore be kept in mind, but the hope of complete restoration can be entertained only if the symptoms recede promptly upon anti-syphilitic treatment. If the symptoms do not in any way yield to treatment, and if, after a number of careful trials the condition remains practically the same, the prognosis is as unfavorable as it would be in any other case of myelitis. The prognosis will also depend very largely upon the evidence of the amount of damage that has been done by the specific process. If the symptoms show that there has been a complete destruction in one or more systems of the spinal cord the possibilities of recovery are naturally less than in cases in which the symptoms point to but a slight involvement of these parts. Yet whatever the outlook may be at the time the patient is examined, it is the physician's duty to warn the parents against the possibility of relapses and to prepare the relatives for the fact that later attacks may involve more vital parts, and may, therefore, be more dangerous than the one through which the child is passing at the time.

TREATMENT.—Syphilis of the spinal cord in the child, as well as in the adult, calls for very prompt treatment. We must depend upon the usual remedies—the mercurials and the iodides. It is wrong to depend upon either one alone, for in some cases the mercurials are more effective than the iodides, and in others the reverse is true. It is my practice invariably to begin treatment with one drug,* and to stop mercury or the iodide if either is not well tolerated or if the improvement is such that I find it unnecessary to push either drug to the extreme. In some instances it is evident that all forms of specific treatment are of no avail. Mercury should be given in the form of inunctions, either of the ten per cent. oleate of mercury or of the unguentum hydrargyri.

* I agree with Lewin that it is best not to give mercurials and iodides in combination.

According to the age of the child, one-half to one gramme of the ointment may be rubbed in daily. If iodides are to be exhibited begin giving small doses of the saturated solution of the iodide of sodium, the daily dose to be increased slowly. Thus I begin with three or four minims of this solution, to be given in milk three times a day, and increase the daily dose by one minim until a child, according to its age, takes ten, fifteen, or twenty minims of the iodide three times a day. As soon as decided improvement takes place the quantity of the mercurial inunction given may be reduced and soon stopped altogether; but the iodides may be pushed for some time after this improvement has been noticed. In cases in which no improvement occurs, in spite of the proper administration of these drugs,* it is well to stop both, for a time at least; but I would advise, under all circumstances, if the diagnosis is safely established, to make repeated trials of these drugs in the manner indicated above; and if a child has passed through several attacks of specific myelitis, it would be well to subject it to periodic treatment by the iodides in the same manner that one would administer treatment for constitutional syphilis. In addition to the anti-syphilitic treatment, the physician will have to prescribe tonics, sufficient exercise, and nutritious diet, according to the needs of the patient. Moreover, in each attack, or as long as the paralysis lasts, it will be necessary to use electricity and massage, as one prescribes them in all other cases of myelitis, whatever the origin of the myelitis may have been.

* Oppenheim advocates the use of iodipin.

CHAPTER XVII.

DISSEMINATED SCLEROSIS.

DISSEMINATED, insular, or multiple cerebro-spinal sclerosis is distinctly a disease of early life. The first pronounced symptoms appear, generally, in the second and third decades of life, but a few prodromata can be traced back to a much earlier period.* The disease, as described by Charcot, is characterized by a very definite set of symptoms; but variations from this type are not infrequent.

SYMPTOMS.—The prodromal symptoms of this disease consist of a weakness of the upper, and sometimes of the lower, extremities, of a slight awkwardness and trembling of the fingers, and of slight subjective sensory disturbances in the arms and legs. The chief symptoms develop gradually, and are present from the early periods of the disease to the very end, but vary much in intensity at different periods of the disease. The most characteristic symptom is the tremor which is observed on voluntary motion of the affected parts. It is not observed when the parts are at rest, and only when a voluntary effort is made; it is spoken of as "intention tremor." The tremor consists of about six to seven oscillations per second (Grasset). It becomes most

* Totzke stated that some of the symptoms were manifested in two cases at birth; in one case at the age of five months, in one at fourteen months, and in two at the age of two years; and of thirty-one cases which the same author analyzed, a very fair proportion began between the fourth and fourteenth years. There is some reason to doubt whether all these cases were typical cases of disseminated sclerosis. In my own practice the youngest child with distinct symptoms of multiple sclerosis was barely eight years of age at the time of observation, and her symptoms did not come on before the tenth year. Marie, who reported three cases of "sclérose en plaques," in infants, in 1883, now states that he would not diagnosticate the cases as such, and believes the "disease very rare in children." Müller, the most recent author (1904), claims that there is no post-mortem evidence to prove the existence of an infantile multiple sclerosis.

distinct if the patient is asked to take hold of any object, to pass a glass of water to the lips, or to take hold of a pencil, as in drawing or writing. At first this tremor is so slight that it does not interfere with the execution of the simpler movements, but as it increases in intensity it becomes a bar to the use of the hands and legs, and may be an annoyance in walking. In attempting to put a glass of water to the lips the water is spilt ("the patient's face and garments are inundated"—Marie), and in writing the tremor is so marked that the pen tears the paper and the writing becomes altogether indistinct.

Next in importance to this tremor is the difficulty in speech. The patient speaks deliberately, slowly, pronouncing each syllable with unusual care (scanning speech), and yet a distinct tremulousness of the voice is apparent. The consonants l, p, g, and r, are most difficult to pronounce. The words in which these letters occur become unintelligible at a very early period of the disease.

Ocular symptoms are very common; we may have, at times, an inequality of the pupils. (Uhthoff noted this in eleven per cent. of his cases.) The reactions to light and during accommodation are diminished, and a myosis is generally present. Nystagmus appears at a very early day, particularly if lateral movement of the eyes is attempted. Upward or downward motion is unaccompanied by this phenomenon. Visual disturbances are not as common as in tabes, but do occur frequently, according to the excellent studies made of this special symptom by Uhthoff some years ago. Clinically we find a narrowing of the field of vision, particularly of color, and a diminution of the actual visual power. The trouble may be unilateral or bilateral. These symptoms are associated with atrophic discoloration of the papillæ, which persists even though an improvement in the subjective symptoms may take place. The eye symptoms are so similar to those observed in hysteria, that on this account alone the two diseases have frequently been mistaken for one another.

A distinct tremor of the tongue and a peculiar vague expression of the countenance are the other head symptoms present in this disease. Charcot described a special facies

of multiple sclerosis, and refers to a vague and uncertain look, the lips hanging down and half open. On the whole there is an expression of mild stupor on the face; this expression is not altogether unnatural, for the mental condition is in many cases abnormal. The memory is weak; there is lack of attention on the part of the patient, who passes easily from laughing to crying, and *vice versa*. Many of the patients are emotional to an extreme degree, and this is particularly true, according to my own experience, of those in whom the disease begins very early in life.

The paralytic symptoms are of a spastic order. The legs and arms are moved awkwardly at first, later with distinct evidence of rigidity. A spastic or spastic-paretic gait is present, and is associated with those other symptoms characteristic of all spastic paralysis and of involvement of the lateral columns of the cord, viz., rigidity of the muscles and increase of the deep reflexes. All these symptoms may for a long time be slight, but as the disease progresses they become more and more intense; the spasticity and weakness grow worse until the patient becomes completely bed-ridden; and the tremor increases in intensity and interferes with the use of the extremities. Speech becomes almost unintelligible, the memory grows weaker and weaker, until a truly pitiable condition is reached. Objective sensory disturbances are not uncommon; Freund has observed them in twenty-nine out of thirty-three cases. In fourteen there was a mere transitory anæsthesia, while in six cases he states that there was a more or less permanent and complete loss of sensation. He also refers to the occurrence of hyperæsthesia, and even to the dissociation of sensation, but this latter form of sensory disturbances is surely a great rarity. Fortunately, or unfortunately for many, the disease covers a long period of years, during which a tolerable condition of health is maintained. In other cases, again, the symptoms described are developed more rapidly, and a few others are added which are not invariably present. This is true of the bulbar symptoms leading to difficulties not only of speech but of deglutition, and occasionally to interference with the respiratory functions.

Paralysis of the ocular muscles, above all of those supplied by the sixth and third nerves, are not infrequent. Vesical trouble has been recorded by Erb and Oppenheim. If we add to the above symptoms the complete paralysis resulting from apoplectiform attacks it will be evident that the disease may become almost protean in its manifestations. Paradoxical contractions and impulsive laughter are frequent symptoms.

FIG. 72.— Degeneration of the Cauda Equina in Multiple Sclerosis. (After Taylor.)

As important as the positive symptoms hitherto recorded are those of a negative order. Atrophy of the muscles, paralysis of the bladder and rectum, and changes in electrical reaction are entirely wanting.

Why such a variety of symptoms occurs in this disease will be understood easily enough when we refer to the morbid anatomy. The course of multiple cerebrospinal sclerosis is slow, and though invariably leading to a fatal issue, death is, in the majority of cases, the result of some intercurrent disease.

ETIOLOGY.— Disseminated sclerosis comes on most frequently after acute infectious diseases, after powerful emotions, and occasionally after severe injuries. Oppenheim has shown that the disease appears after intoxication by metallic poisons, while Marie believes it to be invariably of infectious origin, coming on after typhoid fever, pneumonia, measles, scarlatina, and small-pox.

Heredity is supposed to play an important *rôle* in the etiology of insular sclerosis. In the broader sense it is true; for multiple sclerosis may indeed affect children who are descended from a neurotic stock; but it is extremely rare

to find the disease in both parent and child. The cases of Pelizaeus, quoted approvingly by some authors, are not to my mind typical cases of multiple sclerosis; they belong rather to the hereditary form of spastic paralysis. Krafft Ebing, and others mention refrigeration as a frequent etiological factor.

PATHOLOGICAL ANATOMY.—We can infer from the name that the chief lesions in this disease are irregular sclerotic patches distributed throughout the greater part of the central nervous system. These patches sometimes occur in the brain as well as in the spinal cord; but there is no rule determining their first appearance either in one or the other, and there is, therefore, as little reason for establishing a spinal form or a cerebral form on anatomical as there is on clinical grounds. The plaques have, however, a few fa-

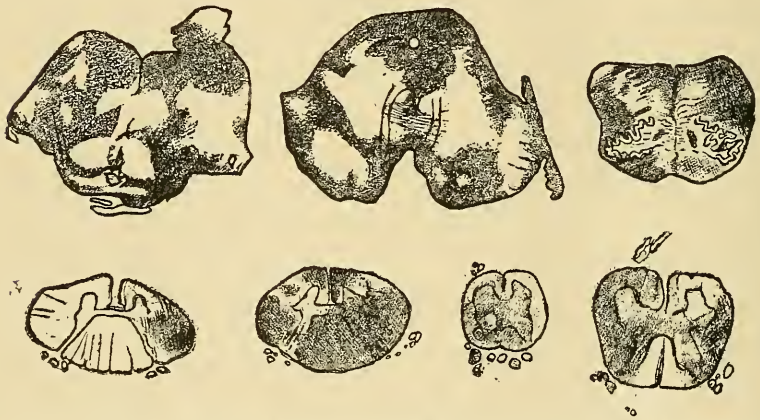


FIG. 73.—Sections through the Pons, Medulla, and Spinal Cord, showing Sclerotic Patches. (Taylor.)

vorite sites; the white matter of the brain, the pons and medulla, the lateral columns and particularly the posterior columns (Ziegler, Müller) in the dorsal and lumber segments; but they also appear in the cortex, in the cervical region, and even in the cauda equina. (Fig. 73.) Charcot was of the opinion that the foci of disease were very rare in the cortex of the hemispheres or of the cerebellum; Taylor claims they may occur in these parts as well as in others. The sclerotic changes may also affect the spinal

roots and various cranial nerves. The optic chiasm is frequently diseased. Whether the sclerosis appears in the central nervous system itself, or in the nerves emerging from it, the character of the changes is very much the same, the main point being that a few, and not necessarily the majority, of the nerve-fibres are destroyed.

These patches present a bluish-gray appearance and are sometimes slightly elevated, at other times the parts in which they occur appear shrunken and contracted. In size they vary from one twenty-fifth to one inch. They are, as a rule, a little harder than the surrounding tissue, and Morris calls especial attention to the nicety with which they can generally be easily differentiated from it by the naked eye.

FIG. 74.—Vicinity of Locus Cœruleus. *c*, cells; *v*, blood-vessels. (Taylor.)

On microscopical examination these patches are found to consist of a dense fibrous tissue, due undoubtedly to the enormous increase of the neuroglia fibres (according to Weigert). Weigert insists that the proliferation of neuroglia tissue is more marked in this disease than in any other. In these sclerotic portions the myelin sheaths, according to the stage of the disease, are more or less diseased, but the axis-cylinders in the majority of the cases remain normal. The entire process may, therefore, be characterized as an interstitial inflammation, and it is owing to the preservation of the axis-cylinder that secondary degeneration in multiple sclerosis is a rare occurrence, indeed; although Buss has recorded a descending degeneration in the lumbar cord and an ascending degeneration of the columns of Goll, and of the direct cerebellar tract from the eighth cervical segment into the medulla oblongata, but his observations are surely exceptional. The absence of secondary degeneration also accounts for the frequent absence of contractures.*

* The tremor of multiple sclerosis has been attributed by some to defective isolation (loss of sheath) of the axis cylinders, on the supposition that the nerve current resembles an electric current, and the axis-cylinder an electric wire; but this is rather fanciful pathology.

Considering the importance which has been attached to the acute infectious diseases as an etiological factor in multiple sclerosis, it would be natural to expect a very early involvement of the blood-vessels in this disease. Such an involvement has been found by Ribbert, Greiff, Cramer, and Marie. Taylor, a more recent author on this subject, whom I am willing to follow in this matter, because the autopsy in his case was on a young subject, proves that in the earlier stages of the disease there is a distinct increase of the smaller blood-vessels and capillaries. (Fig. 74.) The specimens examined also exhibited small hemorrhages and migrating white blood-corpuscles. In some of the vessels there was a slight thickening of the walls and an increase in the number of the nuclei. The perivascular spaces were dilated. The same changes were seen in a second case of Taylor's. In a third case there was absolutely no change in the blood-vessels, but in the second and third cases the disease had lasted longer, and it is, therefore, doubtful whether similar changes in the blood-vessels might not have been present also in these cases in the earlier period of the disease. Taylor thinks that we are not justified in connecting these vascular changes with the sclerotic process, for the patches are not distinctly related to the diseased vessels. Furthermore, vessels are often entirely normal in degenerated areas, and in some cases the disease of the blood-vessels is wholly wanting, however wide-spread the lesions may be.

Strümpell and Müller regard the disease as an endogenous (congenital) affection (say, of the neuroglia), stirred into being by various exciting causes.

Atypical Forms and Differential Diagnosis.—It would be an easy matter to enumerate a large number of atypical forms of multiple sclerosis, for the clinical symptoms as developed by Charcot are not distinct in many cases which have, however, a direct relation to this disease, representing either early forms of the same, or a peculiar localization of the sclerotic patches. One variety of multiple sclerosis with bulbar symptoms bears a close resemblance to amyotrophic lateral sclerosis. But in children the combination of symptoms giving rise to such difficulty in diagnosis is very rare. Then there is the bulbar, or, as Spitzka prefers to call it, "the oblongata type" of multiple sclerosis, in which the usual symptoms are not nearly as well developed as are those pointing to an involvement of the lowest cerebral centres. Difficulties in deglutition, in mastication, and in articulation as well as phonation, are most prominent, and for a long time may obscure the usual symptoms of the disease.

A hemiplegic form of multiple sclerosis has also been observed in the adult, but I have not yet met with it in

children; nor have I seen an apoplectiform beginning of the disease in children, as sometimes happens later in life.

Multiple sclerosis, and forms closely allied to it in children, should not be confounded with transverse myelitis. Acute and chronic forms of myelitis in children are not rare. The acute symptoms and the inflammatory condition upon which they depend disappear promptly enough, but a degeneration is set up which leads to the development of spastic and paralytic symptoms. These might be taken to be symptoms of multiple cerebro-spinal sclerosis unless the history of the patient is carefully considered. The very acute onset and the involvement of the bladder, the persistence of sensory symptoms for a shorter or a longer period of time, will help to differentiate myelitis and its secondary degeneration from insular sclerosis.

From congenital spastic paraplegia the disease can be differentiated by a consideration of the mode of onset, and by the very early appearance of the congenital disease, for insular sclerosis does not generally come on until very nearly the first decade of life is passed. Moreover, in the congenital cases of spastic paraplegia there is frequently some cerebral defect; and the cardinal symptoms of sclerosis are wanting in these cases. Westphal described a pseudo-sclerosis, a condition closely simulating that of multiple sclerosis; but it is not an important disease, for to my knowledge it has not been clearly established in any other instance excepting the one reported by Westphal.

Cases of hereditary tremor might be mistaken for multiple sclerosis, and I have myself seen one patient, a girl aged twelve, in whom there was much doubt as to whether or not this tremor, which became very much aggravated on voluntary motion, was the first symptom of a multiple sclerosis; but having observed the patient for fully five years without the addition of any other symptoms referable to a multiple sclerosis, I am confident that the girl is suffering from an hereditary form of tremor, and have since learned that her father and an uncle have suffered from the same disease since very early childhood.

The question also arises in some cases whether an insular sclerosis or an hysteria is present, but a consideration of the symptoms will soon remove all doubts as to the diagnosis. Nystagmus, scanning speech, and intention tremor are exceedingly rare in hysteria, while the sensory disturbances of hysteria are too typical to be mistaken for the slight subjective and objective disturbances of sensation that occur in multiple sclerosis.

In the adult the chief point in differential diagnosis is considered to be that between multiple sclerosis and paralysis agitans. To this we need pay comparatively little attention, since paralysis agitans is distinctly a disease of later years, and is rare indeed before the age of forty; but I cannot forbear referring to the case of a patient, whom I have

observed many years, who, at the age of fifteen years, presented all the symptoms of genuine paralysis agitans. As will be seen from the accompanying cut, the position of the body, the expression of the face, the position of the hands, are typical of paralysis agitans, yet the symptoms have changed in such a way that at the present time they point to a multiple sclerosis rather than to a shaking palsy. (Fig. 75.)

FIG. 75.—Patient with Paralysis Agitans, who also exhibited some Symptoms of Disseminated Sclerosis.

The patient fell at the age of five years, and had an attack of convulsions with loss of consciousness, which was followed by a slight weakness of the legs, but from this condition he rallied rapidly enough, and was entirely well, exhibiting good physical and mental development. He was bright in school, and perfectly well until the age of fifteen, when he met with another accident; he fell from a carriage and was badly frightened.

The first symptoms he noticed were pains around the left ankle, then a shaking of the left leg. Soon thereafter the arm began to shake. Five months later he noticed the same symptoms on the right side of the body.

On my first examination of the boy, three years after the accident, I found him well nourished; the muscles in good condition, the color of the skin normal, but considerable vasomotor disturbances. He exhibited the mask-like expression of the face, and his speech, as well as the rhythmical tremor of the hands, legs, and head, were exactly those of a patient suffering from paralysis agitans.

Soon another set of symptoms appeared, which proved to me that this "senile" disease, when occurring in a young individual, was subject to peculiar modifications, which allied it much more closely to multiple sclerosis, a disease common in earlier life.

While under observation he developed characteristic nystagmus, scanning,

and tremulous speech. All the reflexes became greatly exaggerated. The typical tremor of paralysis agitans, involving the head, lips, and tongue, and the extremities, remained unchanged. He still exhibits the propulsive movement so characteristic of paralysis agitans. The hands have the position of shaking palsy, and the contractures are much like those seen in the senile form. Motion is almost impossible, and with the symptoms pointing in part to multiple sclerosis, and in part to paralysis agitans, he presents a very unusual appearance.*

PROGNOSIS.—Like many other chronic disorders of the central nervous system, multiple sclerosis is not a rapidly fatal disease. It may run a continuous course for years without seriously endangering life, but any intercurrent disease is likely to lead to a fatal termination. At all events, everyone will acknowledge that it is rare to see cases of multiple sclerosis that have exceeded the age of forty-five or fifty years, and as for those beginning very early in life, the facts are altogether too few to warrant us in stating that such patients live much beyond the middle period of life. The danger to life is greatest in those cases in which the oblongata type predominates. The invasion by disease of the vital centres might lead to early death. The invasion is, however, a very gradual one, and sudden death need not be looked for. In those cases in which such a termination is threatened, periods of asphyxia, of aphonia, and other symptoms of vagus disturbance would occur, and would give warning of the terminal possibilities.

The disease is absolutely incurable, although remissions occur in which all the symptoms excepting the tremor or the nystagmus disappear, and in some cases the disease may come to a complete standstill for a period of several years. In this respect multiple sclerosis is not unlike tabes. My notes of the cases beginning in early life would go to show that the disease is even more slowly progressive than it is in the most favorable adult cases.

TREATMENT.—Keeping in mind what was said relative to the prognosis of the disease little can be expected from our attempts at treatment. But if we cannot cure the disease we can at least secure greater comfort for the patient. As the tremor is most marked when the patient is ac-

* Schultze has reported a similar case in Virchow's Archiv, Bd. LXVIII.

tive—and the disease is exhausting at best—it is wise to keep the patient in bed for a prolonged period of time. The rest cure I have found as efficient in these chronic organic diseases of the central nervous system as I have in the treatment of purely functional troubles. It is often surprising to note how much benefit, and, above all, how much comfort, patients derive from a complete rest when they have been attempting to go about from place to place, often from physician to physician, with their weak and shaking limbs.

With this rest treatment I am in the habit of combining mild hydro-therapeutic procedures. Generally a tepid bath in the morning, followed by lukewarm and successively colder douches down the spine, or by the use of the drip-sheet. At night it is well to give a tepid bath without douches, as the former contributes much to the sleep and the general contentment of the patient. The galvanic current may be employed to advantage. Stabile currents of 10 to 15 milliampères may be passed down the spine and through the extremities. The faradic current should, however, be avoided, as it tends to increase the spastic rigidity of the muscles. Massage is also in order in these cases, if intelligently adimnistered. Spastic rigidities yield a little to such manipulations, and according to careful investigations of Mitchell and his son, the general condition of the patient is improved by massage. If treatment is directed in this way and adapted to each individual case it will be far more satisfactory than sending the patient away to various springs. The Frenkel method of exercises, that has proved itself to be of great value in tabes, is certain to be helpful in these cases, if intelligently applied.

Various drugs have been employed and recommended in cases of multiple sclerosis. Among these nitrate of silver, the iodides, and mercurials are the most prominent. I am confident that the first never does any good excepting possibly to allay pain, and the last two will not be needed, for it is very certain that multiple sclerosis is rarely developed on a syphilitic basis, and there is no indication for the use of resorbents.

CHAPTER XVIII.

COMPRESSION OF THE SPINAL CORD—POTT'S PARALYSIS.

Injury to the cord may be due to specific diseases in the bone, to compression by an aneurism or to growths originating in the membranes. These conditions are very rare, however, in children, and whenever the symptoms point to compression of the cord, caries of the bone may be taken to be the primary trouble. Pott's disease of the spinal column begins so insidiously that the recognition of it and of the symptoms resulting from compression are reached frequently by exclusion only. While the disease may develop at any period, it is more common in childhood and in early life; it is the result of a tubercular or scrofulous diathesis; and evidence is easily adduced in the majority of cases of the existence of such diathesis in the same family and in the same or in preceding generations. The slightest traumatism may be sufficient to bring about marked disease of the bone, but evidently only in those children who are predisposed to such disease, so that here as elswhere the traumatic incident simply helps to bring out the latent tendency to disease. There is often a period of weeks or months between the time of injury and the first symptoms of the disease. In consequence of the caries of the bone, the bodies of the vertebræ become softened and are displaced, two or three of them, or even more, forming a marked angular curvature. Such angular curvature may in rare cases be the result of a septic process, in which case an abscess may have formed in the bone and caused the softening. Generally the bodies of the vertebræ become diseased and the intervertebral substance undergoes inflammatory changes as well. Vertebral disease may lead to retropharyngeal, mediastinal and psoas abscesses.

The deformity resulting from the disease of the bone is of such a character that a sharp angle is usually formed at one point, at which the cord is generally much compressed. In some instances the cord becomes quite thread-like. It is remarkable that the cord may sustain a very large amount of compression before its functions are seriously interfered with, or the symptoms of disturbed function appear. This accounts for the fact that in many instances the angular curvature may be extreme and yet neither paralysis nor anæsthesia is observed during life. Lateral displacement may occur without its having any influence upon the cord. Very naturally the spinal cord suffers under these conditions, and the roots emerging or entering through the intervertebral foramina are compressed by the thickened sheaths or by other inflammatory products. This accounts for the frequent occurrence of pain in these cases, and for the occurrence of paralysis in cases in which the cord itself shows tolerably normal structure.

The causes of injury to the spinal cord in the subjects of Pott's disease are twofold: first, the cord is actually compressed in many cases, and, secondly, it suffers from the inflammatory products gathering between the cord and the bony parts. The latter cause is often quite as active as the former. The changes in the cord itself will vary very largely according to the degree of compression. The cord is somewhat altered, the normal white having given way to a grayish or reddish-gray tint. The cord is less hard than under normal conditions, both these changes being clearly due to inflammatory processes. It has been found by Schmauss and others that the amount of inflammation of the cord is not always in keeping with the degree of compression. Recent French authors have attached considerable importance to an œdema of the cord produced by compression of the veins as they issue from the cord. The œdema is subject to fluctuations; and a diminution of the same may be responsible for an improvement in the paralysis.

The symptoms of Pott's paralysis often set in long before there is any possibility of actual compression. In those cases we must suppose that the symptoms are due to a

pachymeningitis constricting the spinal roots and blood-vessels; the occurrence of rapidly developing spinal abscess, associated with painful paralysis, and the disappearance of the paralysis as soon as the abscess is emptied, lend support to this view. By degrees an extension of the inflammatory process takes place into the substance of the cord. The myelitis as it occurs with Pott's disease shows a general increase in the interstitial tissue, with proliferation of the various cells and degenerative changes in the more important nerve elements. Granular corpuscles, corpora amylacea, and globular masses of myelin are found in sections that are properly hardened. The gray matter may have become almost indistinguishable from the white, and the larger ganglion cells may have entirely disappeared from the former. In addition to these changes the usual ascending and descending degenerations will occur, as from any other focus of inflammation in the cord. The changes in the peripheral nerve-roots will be those that would be expected from actual compression and inflammation. The interstitial tissue will be increased, the nerve-fibres themselves will be wasted, and the axis-cylinders will show the usual changes of degeneration.

Symptoms.—The chief symptoms in this disease are a very gradual development of a spastic paralysis, generally of the legs and possibly of arms and legs; the reflexes are increased, and radiating pains are present at a very early period of the disease. The symptoms often resemble those of a dorsal myelitis, since Pott's disease attacks the dorsal region more frequently than any other. If other regions of the spine are diseased the symptoms will vary as they would in the various forms of myelitis, but root symptoms will always play a very prominent part. The symptoms due to compression of the root-fibres will also help to differentiate between a myelitis following Pott's disease and the ordinary traumatic or non-traumatic myelitis. The compression of the sensory nerve-roots causes severe pains. It may also interfere with normal sensation. (Dissociated sensation has been noticed in several of my cases); anæsthesia dolorosa may be associated with the paralysis. Before the condition of anæsthesia is established a transitory

hyperæsthesia may have existed. The irritation of the motor root-fibres would lead us to expect a condition of involuntary contractions of the muscles, but, as a matter of fact, these are not so frequent, and probably because the ventral gray matter of the cord, being nearest the vertebral bodies, is affected at an early day; and since the function of this gray matter is impaired, irritation of the anterior roots connected with it will not be able to exhibit symptoms of irritation. According to the location of the disease in the bone the symptoms may for a time remain unilateral; but, as a rule, the disease spreads rapidly enough to bring about a bilateral, and often symmetrical, set of symptoms within a very short period of time. Furthermore, as the disease progresses the paralyzed parts may become atrophied, the vesical and rectal reflexes may be impaired, and we then have all the ordinary symptoms of myelitis, even including bed-sores and the usual trophic disturbances of the skin.

The course of the disease may be inferred from the rapidity with which all these symptoms are developed. If the disease is of a slowly progressive character, the symptoms will naturally be developed one by one, whereas if the bone disturbance is of a more pernicious and rapidly progressive character the symptoms of a complete myelitis will be reached at a very early day.*

Diagnosis.—Paralysis due to Pott's disease must be distinguished from subacute or chronic myelitis. This distinction is not always an easy one to make. It is reached most readily in those cases in which the course of the disease is a very slow one and the usual causes leading to a transverse myelitis are wanting. It is rendered probable by the coexistence of pain with the paralysis, and above all things by the discovery of tenderness to pressure over the spinous processes. These same symptoms might occur in cases of tumor of the cord, but the unilateral development of symptoms in the latter disease will soon lead to a safe differential diagnosis. (See next chapter.) The earlier symptoms of a Pott's paralysis may simulate the condition

* In some cases, like the one reported by Daxenberger, the caries may be latent for years, before spinal-cord symptoms are developed.

of spastic paraplegia, either of the congenital order or of that form due to specific disease.

In several cases, under the author's care, the symptoms of Pott's paralysis were developed long before any actual evidence of disease of the vertebræ could be elicited. Whitman also states that in a number of cases the paralysis precedes the deformity. The diagnosis must at times be reached by exclusion; but the condition may be suspected in cases of a gradually developing spastic and painful paralysis, in which there is no trace of preceding injury, and no history of any acute infectious disease immediately preceding the onset of the symptoms.

The age of the patient and his general appearance will also help to establish a differential diagnosis.

PROGNOSIS.—The progress of the paralysis will depend mainly upon the development of the disease in the bone. If the disease has lasted long enough to produce complete compression at one point, the spinal-cord symptoms may persist during life, although the bone disease be checked or recovered from. In other cases the course of the spinal-cord disease may be checked, although the disease of the bone steadily progresses. With the subsidence of the inflammation the sensory symptoms may diminish, but the condition of paraplegia may continue years thereafter as a natural result of the changes that have been set up in the spinal cord. In a number of cases, however, there is a proportionate development between the disease of the bone and the disease in the spinal cord. In the more severe cases the paralysis remains unaltered, or is steadily progressive, and all those complications arise, such as bed-sores, cystitis, and the like, which help to bring about a fatal issue in many cases of myelitis. In the milder cases recovery is to be expected, and many a child who has not been able to walk for years during the active period of caries has attained a tolerably normal gait in later life. On the whole, the motor symptoms are much more persistent than the sensory. The prognosis is much more grave in the cases of cervical Pott's disease than in those occurring in the lower vertebral regions, for reasons which can be readily understood.

TREATMENT.—In the treatment of paralysis resulting from Pott's disease surgical methods should be resorted to

at the very beginning. If the spinal cord is to recover, the tubercular process in the spinal column must be checked first of all. The old method of absolute rest should be employed, and tonics, such as cod-liver oil and iron, should be given. I do not wish to enter into the discussion as to which of these measures, the rest or the tonics, is more important. Some will be cured by the one, and some by the other, method, but I have not the slightest doubt that if all these measures are employed conjointly the result will be more satisfactory. In addition to rest, absolute immobility should be secured, either by forcible extension in bed, or by the application of a plaster-of-Paris jacket. The plaster-of-Paris jacket is not so useful during the period of rest as it is during the period of partial recovery, in which it is desirable for many reasons that the child should be taken out of bed and enabled to walk in the open air. I am very certain that the application of counter-irritants, which is still recommended by many, is of no use, and simply helps to increase the tortures that these children suffer. Of late years active surgical interference has been suggested in these cases by Macewen, Horsley, and others. Thorburn and Chipault's statistics show a mortality of forty-three per cent. The surgeon's skill should, at all events, be restricted to those cases in which the clinical symptoms prove that there is evidence of compression at any one special point by inflammatory products or by displaced bone. If the child is strong enough to tolerate such surgical procedures, the removal of these products of inflammation may be attempted; but it is well to caution the physician who may be eager to try new surgical procedures that children are less tolerant than adults of spinal and cerebral operations. Callot's redressement forcée cannot be endorsed.

The orthopedic surgeon, by well planned tenotomies, may enable the child to use its feet to greater advantage. His treatment will be supported in every case by the discreet use of electricity and massage. The ordinary blood and nerve tonics will help very much more than the iodides or mercurials, but these should be given in the earliest stages of the trouble. I have seen favorable results from mercurials in cases in which the process was assumed to be gummatous and not tubercular.

CHAPTER XIX.

TUMORS OF THE SPINAL CORD AND ITS MENINGES.

THE study of tumors of the spinal cord was at one time considered to be of small practical value on account of the rarity of the affection; but far greater interest attaches to it now from the fact that the removal of these tumors has been rendered possible by the recent advances in surgery. About fifteen years ago Mills and Lloyd collected and analyzed fifty cases of cord tumors; fourteen per cent. of these were under twenty years of age. Bruns, Oppenheim, Schlesinger, and others speak of the relative frequency of occurrence of these tumors in the early years of life.

CAUSES.—The actual cause of tumors of the spinal cord can be determined as little as in the case of cerebral tumors. They are rarely primary, and are commonly due to some constitutional diathesis, such as tuberculosis or syphilis, or are the metastatic products of malignant tumors in other organs of the body. The tendency to proliferation of nerve-tissue is not so marked in the spinal cord as in the brain of the child. Gliomata originate in the vicinity of the central canal, and here are of special interest, as they often break down and lead to the formation of cavities, and hence have given rise to the discussion whether these cavities are invariably the result of such disorganization of a neoplasm, or whether the condition of syringo-myelia may be the result of congenital enlargement of the central canal. Small hemorrhages and embolism, though much rarer than in the brain, may give rise to cystic formations, but among cystic growths those due to parasitic infection, particularly to the echinococcus, are the most common. As in the case of brain tumor, accidental injury has been declared responsible by many for the occurrence of spinal tumors. It is doubtful, however, whether traumatism plays any further *rôle* than that either of eliciting a latent tendency to such disease, or of merely calling attention to the symptoms of the disease which may have antedated the injury.

SYMPTOMS.—The symptoms of tumor of the cord might be deduced from a consideration of the disturbance of

function due to a very gradual invasion of the cord. Thus in many cases the symptoms are strictly unilateral for a time, and become bilateral with the extension of the disease from one part of the cord to the other. The symptoms will naturally vary also according to the seat of the disease, according to the more or less rapid destruction of the cord, and according to the development of the tumor in the meninges or in the spinal cord itself. It is of the greatest importance to distinguish between extra-medullary and medullary tumors. Extra-medullary tumors are characterized by the early appearance of root symptoms, more persistent pain in the back, and by sensory disturbances which point to compression of the posterior root-fibres; by paralysis and atrophy if the growth is on the ventral surface. These distinctions will not hold good if, as in a case of my own, the intra-medullary tumor starts near the entrance of the posterior root-fibres and actually compresses these. In some cases, however, the disproportion between the sensory symptoms and the paralysis will direct attention to the seat of the tumor outside of the spinal cord, particularly if all symptoms pointing to involvement of the central gray matter are absent. The focal symptoms in extra medullary, as well as in intra-medullary tumors will vary greatly according to the seat of the tumor. There may be but slight sensory disturbances in a single area or in one limb; the sensory disturbances may be unilateral, and these may or may not be associated with partial paralysis, either unilateral or bilateral; or there may be crossed sensory and motor symptoms. Any set of spinal, and more particularly hemi-spinal symptoms, whether of a motor, sensory, or vasomotor order, if they be slowly progressive, are suspicious of tumor. There will be additional reason to suspect tumor if there is evidence of a distinct constitutional diathesis. All the symptoms of various spinal-cord affections may be simulated by tumor, according to the part of the spinal cord affected, and in this respect the symptomatology of tumors may be quite as varied as that of syphilis of the spinal cord.

If we endeavor to analyze the symptoms in detail, we find that disturbances of sensation are by far the most com-

mon. If the tumor is in the cervical region, the pain radiates into the upper extremities, and generally into definite parts of the brachial plexus; if in the dorsal region, the pain is relegated to the region of the ribs, the thorax, or the abdomen; if in the lumbar region, the pain is described as limited to the various parts of the lumbar or sacral plexus. Whatever the level of the lesion may be, the sensory disturbances may be unilateral for a time and gradually become bilateral. Vasomotor disturbances occur, similar to those in peripheral neuritis; the skin becomes glossy, livid, and painful. With these symptoms muscular wasting and paralysis will soon be associated. As the disease progresses the hyperalgesia or the paræsthesia gives way to complete loss of sensation, the paralysis becomes complete, and the vasomotor disturbances increase, and serious trophic disturbances may set in. The sensory changes may vary in still another respect. Since the tumor need not invade all sensory fibres at once, some may remain entirely intact, while the function of others becomes seriously impaired. Thus dissociated sensation may be present in some cases, but is rarely as distinctly developed as in the case of syringomyelia, which, as we have seen above, may or may not be connected with the formation of a central neoplasm. According to well-known physiological laws, and to the old-time teachings of Brown-Séquard, one would expect in many of these cases of tumor to find distinct symptoms of unilateral lesion of the cord, namely, paralysis in one member or in one-half of the body, with loss of sensation in the opposite half; but this, curiously enough, is not often the case.

The paralysis will be developed in accordance with the ordinary laws relating to spinal-cord affections. The muscles represented in the segment or segments involved by the neoplasm (see tables in Chapter I.) will not only be paralyzed, but will also undergo atrophy, while the muscles represented in lower segments will show spastic forms of paralysis without any atrophy. The changes in electrical reactions will also vary in accordance with these conditions, the only muscles exhibiting distinct reaction of degeneration being those governed by cells in the affected

segments. Spasmodic contractions, the ordinary rigidities and contractures, will be observed in these cases under the same conditions as in cases of myelitis. All the symptoms will be largely influenced by the fact that an active myelitic process is started up sooner or later in the vicinity of most of the tumors. This is particularly true of tubercular and gummatous deposits, whereas other forms of tumor may exist for some period of time without seriously affecting the tissue in their vicinity.

Paralysis of the sphincters of the bladder and rectum will occur in cases of tumor as in cases of myelitis. In the case of tumor in the lumbar region, absolute paralysis of the sphincters and retention are the natural results, while if the tumor is above this level incontinence will be the more frequent condition; but in the later stages retention with overflow is the rule. The conditions of the sphincter ani will vary in very much the same way. Tumors of the cauda equina deserve especial consideration. In addition to the general symptoms, pains radiating into the sacrum, the perineum, the anus, the bladder, and into the distribution of one or both sciatic nerves will be present. In one of my own cases, bladder and rectal symptoms were absent for a long time. The gait may be only slightly defective for some weeks; with the growth of the neoplasm the paralysis of the legs will become more and more evident. In two patients whose cases were reported by the author, pressure to one side of the median line had produced the precise sensory disturbances of which they had complained.

It is well to remember that a very large number of so-called "spinal cord tumors" start from the vertebral bodies; many others from the meninges, and that the spinal symptoms are in reality compression-symptoms, and not due to an invasion of the cord. Not infrequently in children, as I had occasion recently to see in a lad of fourteen years, the spinal tumors are so clearly metastatic products (sarcomata) that operative interference would be of no avail. In some instances of tuberculosis, spinal symptoms may appear to be primary; but the autopsy will reveal the more probable focus of infection. In a case of solitary tubercle of the cord between the sixth and seventh cervical segments I found in addition that the cord between the seventh cervical and the fourth dorsal segments was a diffluent mass. There were also minute tubercular deposits discovered in both lungs, and in both apices there were incipient cavernous spaces.

PATHOLOGY.—The morbid anatomy of tumors of the cord differs very little from that of tumors of the brain. The central nervous system is in this respect a unit, and the same growths which are apt to occur in the brain occur, though with lesser frequency, in the spinal cord. In the cases of Mills and Lloyd, only four were of a tubercular character, while twelve were either sarcomata or gliomata. Parasitic growths occurred in three cases of the fifty, syphilitic growths in five, and the remainder were distributed among various kinds of neoplasms, such as myxoma, fibroma, carcinoma, and psammoma. Neuromata of the cauda equina must be included in the list. The largest collection of cases of solitary tubercle of the spinal cord has been made and carefully analyzed by Dr. Herter. His experience proves that the disease is one of early life, twenty of the twenty-six cases occurring before the age of thirty-five. A few authors have reported cases of tubercle which they supposed to be primary; but, as in the case reported by myself, although it was difficult to prove any other focus of tubercular disease during life, such focus of disease evidently precedes the spinal disease in every instance. Specific infiltration of the spinal cord is, on the whole, more frequent than the same infiltration of the brain. To be sure, relatively few cases of this description are on record, but I am certain that more careful examination will hereafter reveal a larger number of gummatous deposits, if not gummatous growths, in different parts of the cord, associated with the specific meningo-myelitis, which is, as has been stated in another chapter, the most frequent form of syphilitic disease of the cord. Tumors of the cord may occasionally erode the surrounding bony parts and may work their way to the surface, but this is very much rarer indeed than a similar invasion of the skull in cases of brain tumor. In a case of my own the tumor (though a small one) "presented" though an intravertebral foramen.

DIAGNOSIS.—Tumors of the spinal cord are to be differentiated from hemorrhage into the cord, from spinal caries with Pott's paralysis, from pachymeningitis cervicalis, transverse myelitis, syringo-myelia, neuritis, and from injuries of the cord.

In the case of hemorrhage the paralysis and other symptoms resulting

therefrom appear with great suddeness. The condition is one of extreme rarity, and in many of the cases leads to complete paralysis, with all its complications, and to death in a relatively short period of time. A confusion can, therefore, exist only in the first few days of the disease, and under such circumstances the suddenness of the onset would argue strongly in favor of hemorrhage.

Spinal caries, with the resulting paralysis, may occasionally present symptoms similar to those of spinal tumor. In both, pressure symptoms are present, but in the case of caries of the bone the symptoms are not nearly so intense, as a rule, as in those due to the presence of neoplasm. The chief reliance in differential diagnosis must, however, be placed upon the detection of bone trouble in the case of spinal caries, although, as we have seen, such evidence does not always precede the onset of pressure symptoms. The more rapid course of the symptoms in spinal tumor, and the negative results of treatment, will also help to differentiate the two conditions. In caries of the vertebræ, symptoms pointing to the ventral (anterior) surface are a little more frequent than in tumor.

As between pachymengitis cervicalis and tumor of the cervical region, there are but few points of differential diagnosis, and the diagnosis will have to rest very largely upon the excessive rigidity, the very typical distribution of the paralysis in the hand, and the more acute pain in cases of pachymeningitis than in those of tumor. In tumor, moreover, the symptoms are more apt to be unilateral for a time, while those of pachymeningitis are, as a rule, bilateral and symmetrical.

Some of these same points of differential diagnosis will help us also to distinguish between transverse myelitis and tumor of the cord, the chief points of difference being these: In myelitis the onset is much more rapid than in tumor, there is less pain, and the entire cross-section of the cord shows symptoms of involvement very much more promptly in myelitis than in the majority of cases of neoplasm. A doubt as to which of these two conditions is present can exist in the first few days or weeks only; after this the general course of the symptoms will determine the character of the morbid process.

Syringo-myelia is closely related to tumor of the cord; there is doubt, in fact, whether some, if not many, of the cases of syringo-myelia do not represent primary neoplasm of the cord, with secondary enlargement of the central canal and a breaking down of the spinal tissues. But the gliosis which may or may not be the starting-point of a syringo-myelia, takes an extremely chronic course, and it is, therefore, by this fact alone, as well as by others, that the two diseases can be sufficiently distinguished from one another. In syringo-myelia the symptoms are often strictly bilateral, or symmetrical, and the disturbances of sensation (the loss of pain and temperature sense, while the tactile and muscular sense are preserved) are very typical of syringo-myelia and are rarely met with in cases of spinal tumor. In syringo-myelia there is also evidence in many cases of a slowly ascending or descending morbid process and such slow ascent or descent is not common in cases of tumor. The differential diagnosis is one of great importance, for the one

disease is compatible with prolongation of life, while the other is rapidly destructive. (See end of this chapter.)

The symptoms of neuritis may for a time obscure those due to tumor. In cases which I have observed the first symptoms that appeared were those of a neuritis involving various branches of the brachial plexus. These were due to compression of the spinal roots, and are naturally present in those cases in which the tumor begins either in the meninges or near the surface of the cord. The early appearance of complete paralysis and the development of other symptoms, particularly those pointing to interference with the vesical and rectal reflexes, will argue in favor of neoplasm and not of a simple neuritis. The rapid progress of the symptoms in the case of tumor, the entire absence of fever in most of them, will also point to tumor rather than to neuritis. The presence of sensory and paralytic symptoms in both halves of the body, or in the lower as well as upper extremities, without involvement of the bladder or rectum, may be considered as pointing to a multiple neuritis rather than to tumor.

A localized specific meningitis may at times simulate tumors of the cord, and, as was indicated before, the frequent association of gummatous deposits with such specific meningo-myelitis may present unusual difficulties of differential diagnosis. In one case under my observation for a period of several months, it was impossible to differentiate accurately between these two conditions; and unfortunately the result of treatment is not to be depended upon, for even in cases of a distinctly specific character the symptoms do not always yield to antisyphilitic treatment.

Severe injuries of the spinal cord may present symptoms resembling those of tumor. But the history of the case, and the extreme painfulness over the spinous processes, will prevent the possibility of confusion between the two.

PROGNOSIS. — In spite of increasing surgical successes tumor of the cord is an extremely grave disease. Spinal growths are secondary to malignant disease in other parts of the organism and help to intensify the gravity of the disease elsewhere. In the case of tubercle, sarcomata, and carcinomata the prognosis is absolutely bad, while it is only a little better in cases of gummata.

The duration of the disease varies according to the site of the tumor and according to the morbid character of the new growth. Tumors of the cervical region are, on the whole, more rapidly fatal than those in the dorsal or lumbar region. Tubercles, carcinomata, and sarcomata, take a more rapid course, as a rule, than the other forms of neoplasm.

TREATMENT. — Tumors of the spinal cord and its meninges cannot be sensibly affected by drugs. There is a

possible exception in the case of gummata; yet, as was intimated above, the effect of specific treatment in these cases is, to say the least, extremely doubtful. I would advise, however, that every case of tumor of the cord be given the benefit of the doubt, and that active specific treatment by iodides and mercurial inunctions be attempted. If these drugs avail little, no other drug need be exhibited.

Much has been expected of surgical treatment; contrary to our experience with brain tumors, these expectations are nearer fulfilment. Extra-medullary growths are the only ones that we can expect the surgeon to remove. Success has been reported by Gowers, Horsley, Starr, Bruns, Oppenheim, Spiller, myself, and others.* The cases most favorable for operation are those which occur near the surface of the cord or in the meninges, and those in which the operation is done at a relatively early day, before the neoplasm has invaded the cord or has been followed by secondary myelitis and secondary degenerations. The cases published by Laqueur, in which the tumor was found outside of the dura mater and compressing the cauda equina, those reported by Oppenheim, Dejerine, Fraenkel, Putnam, Boettiger-Krause, and myself, make it almost incumbent upon us to urge operation in extra-spinal tumors.

The question arises whether extirpation of the tumor should be attempted in cases of a tubercular nature and in those in which gumma is suspected. In tubercle of the cord, even the successful removal would in all probability prolong life but very little, so that surgical interference is scarcely warranted; and yet in a case that is otherwise favorable for operation there seems to me to be sufficient reason to interfere, if there are no other symptoms of general tuberculosis. I am in favor of removing gummata if the tumor has resisted, as it generally does, every form of treatment, provided such treatment has been carried out religiously for a period of from three to four weeks without any sign of improvement. I consider Horsley's advice entirely sound, that if a tumor has resisted every form of treatment for a sufficient length of time, and if the conditions are

* Oppenheim states that in 17 of 46 tabulated cases (36 per cent.) cure or improvement was obtained as a result of surgical interference.

favorable for surgical interference, the proper surgical procedures should be resorted to, and that, too, without much delay, for delay is far more serious under these circumstances than prompt and proper surgical interference would be.

SYRINGOMYELIA AND GLIOSIS OF THE CORD.

Syringomyelia, or, more properly speaking, "myelosyringosis," is a form of disease that is rare enough in the adult, and still rarer in children. Its symptoms resemble somewhat those of tumor of the cord, of amyotrophic lateral sclerosis and of progressive muscular atrophy. As the name indicates, the disease is due to cavity formation in the spinal cord. This may be the result of a congenital abnormality of structure, or it may be due to a gliomatous process starting in the vicinity of the central canal, and causing a destruction of tissue. (See monographs by Hoffmann and Schlesinger.)

We need not discuss whether the one or the other mode of origin is the more frequent, and on this point opinions still vary. The cavity is, as a rule, largest in the cervical segments, but may be continued up into the medulla and downward into the dorsal and lumbar segments.

SYMPTOMS.—Less than fifteen years ago syringomyelia was considered a mere curiosity without any special practical importance; but the investigations of Schultze and of Kahler have enabled us to recognize the disease by a very definite set of symptoms. The symptoms will naturally vary according to the extent of the cavity; but as the cavity is generally most developed, and has its beginning in the cervical region, the symptoms are first observed in the upper extremity and generally around the shoulder.

We may divide the symptoms into three groups: The first group consists of trophic disturbances, affecting the skin, its subcutaneous tissues, and the bones. Glossiness of skin, particularly of the fingers, deep fissures, felons—which are sometimes painless—and phalangeal necrosis, with marked distortion of the fingers, are the most frequent. Patients also complain of burning, prickling feelings, and of a sensation of numbness. These various disturbances are associated at an early date with the second group of symptoms, consisting of partial disturbances of sensation. There is either a diminution or a complete abolition of the sense of pain and of temperature, while the sense of touch and of muscular innervation remains absolutely normal.

The third group of symptoms includes a progressive atrophy of the muscles, beginning in the small muscles of the hand, and gradually involving the forearm, the arm, and the shoulder muscles. The resemblancce to the Aran-Duchenne type of progressive muscular atrophy is very great (see Chapter XXII), and it is only by the association of this progressive muscular atrophy with dissociated disturbances of sensation, and with the trophic symptoms described above, that we can differentiate between syringomyelia and progressive muscular atrophy, as well as amyotrophic lateral sclerosis. In progressive muscular atrophy of the Aran-Duchenne type, there are no marked disturbances in the skin and subcutaneous tissue; and in amyotrophic lateral scle-

rosis there are no disturbances of sensation, and no marked trophic symptoms, but in this disease and in syringomyelia the reflexes may be exaggerated.

In syringomyelia other symptoms occur, which will depend largely upon the extent of the cavity formation. If the cavity encroaches very largely upon the anterior gray matter, there will be a large amount of flaccid atrophy and paralysis, the electric reactions will be disturbed, and the reflexes will be

FIG. 76.—Section through Seventh Cervical Segment, showing Gliosis of Cord. (Herter.)*

diminished in the parts governed by the affected region. Fibrillary tremor is present in some cases. All these symptoms are generally most pronounced in the upper extremities, since the cavity is greatest in the cervical region, and if the lateral columns in the cervical region are involved, we may expect a spastic paralysis of the lower extremities with increased reflexes, rigidity, and contractures. Cutaneous reflexes are, at times, normal, at other times diminished or entirely absent. Hyperidrosis or hypidrosis (unilateral or bilateral) have been reported. In the cervical cases a narrowing of the palpebral fissure and

* This and the following figure are reproductions of specimens prepared and photographed by Dr. Herter, and kindly furnished me for use in this book.

contraction or dilatation of the pupils must be added to the possible symptoms of this curious disease. If the affection extends into the medulla and pons, disturbed sensation in the area supplied by the trigeminus is present, together with atrophy and paralysis of the tongue, hoarseness, difficulty of deglutition, and disturbances of the senses of taste and hearing. Other cranial nerve nuclei may be involved; nystagmus may be present; polyuria and increased salivation have also been noticed. Pains and rigidity in the upper portion of

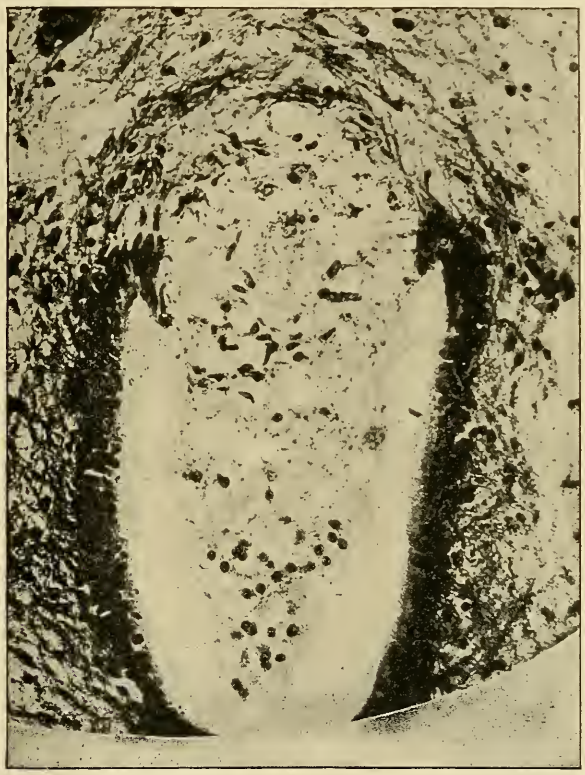


FIG. 77.—Section through Part of Fourth Dorsal Segment. The central canal is invaded by glia cells and surrounded by groups of cells and nuclei resembling those of the ependyma. (Herter.)

the spinal column are not infrequent, and scoliosis or a combination of scoliosis and kyphosis may also be present. Vesical and rectal disturbances are at times superadded.

Syringomyelia is practically a disease of adolescence and of adult life, but some of its symptoms can at times be traced back to the second decade of life. The earliest case which I have seen was in a woman of twenty, in whom the first symptoms began at the age of fifteen. It is interesting to note, however, in this connection, that a disease which is intimately related

(at least in its pathology) to syringomyelia, has been observed by Dr. Herter in a child one year of age. Through his kindness I am enabled to insert a summary of his (unpublished) case, which he reported to the New York Neurological Society.

Three months before the child was examined the right arm and hand began to be weak, and the head drooped forward. The examination brought out the following points: Complete loss of power and atrophy of muscles in both arms and shoulders; the muscles were flabby; there was slight movement of hands and fingers; rigidity of neck; the knee-jerks were increased, especially on the right side; ankle clonus was elicited on both sides; there was an irregular rise of temperature, ranging between 100° and 105° F. daily. The child died two weeks after examination.

The post-mortem examination revealed considerable enlargement of the right half of the medulla oblongata and of the cervical cord extending to the eighth cervical segment. There was some enlargement from this point to the sixth dorsal; below this the cord appeared to be normal. In the upper cervical segments the nervous structures of the spinal cord were replaced by a new growth; in the lower cervical region the pressure effects of the new growth were evident. At about the level of the seventh cervical segment the central canal was lined with an unbroken layer of columnar ependymal cells, except at the posterior wall, which was broken through by a mass of larger glia cells, which partially filled the canal. (Fig. 77.) The glioma invaded nearly the entire cross-section of the cord, but undoubtedly originated in the vicinity of the central canal.

The case is not unlike one reported by K. Miura; Sokoloff described a glioma of the medulla, with cavity formation, in a boy, aged five years.

CHAPTER XX.

FAMILY DISEASES OF THE SPINAL OR CEREBRO-SPINAL SYSTEM.

FAMILY affections of the entire central nervous system constitute an important group of diseases. They are the result of defective development, or possibly of early degeneration of various parts and tracts in the brain and spinal cord. Of these disorders hereditary ataxy, or Friedreich's disease, is the best known. A few years ago Nonne and Marie described a type of disease which bears a close resemblance to hereditary ataxy, and for which Marie proposed the term Hereditary cerebellar ataxy (Heredo-ataxie cerebelleuse). In Friedreich's disease the morbid process is restricted largely to the posterior or to the postero-lateral columns; in Marie's disease there is in all probability a defective development of the cerebellum and of the tracts connected with it. Congenital or hereditary defect of the pyramidal tracts also leads to another distinct group of diseases which we may term Hereditary spastic paralysis; and Hoffman has still more recently described an hereditary form of progressive muscular atrophy of spinal origin in which the chief defect was found in the anterior gray matter of the cord. These hereditary diseases suggest a clinical relationship to syphilitic disease of the spinal cord (which may be inherited, though not hereditary) and to some forms of cerebral birth palsies. Moreover, some of these hereditary affections are distinctly cerebro-spinal, both as regards the distribution of the lesions and the character of the symptoms. It is no easy task to separate these various forms from one another and to bring order out of chaos. The family form of progressive muscular atrophy will be considered to much greater advantage in connection with the other forms of progressive muscular disease.

HEREDITARY ATAXY.—FRIEDREICH'S DISEASE.

Hereditary ataxy, or Friedreich's disease, as it is called in honor of the physician who gave the first satisfactory account of it, is a family affection. Several generations are often affected by it; but naturally we may find every now and then that the disease occurs in one or several members of a family without any history of a similar trouble in the patient's ancestry. It is not necessary, therefore, to speak of such cases as non-hereditary forms of Friedreich's ataxia, as an English writer has recently done, for the disease must invariably have a starting-point, and the patients under observation may indeed be the first of a long series that are to follow. The children who have come under my special notice in three different families gave no history of any similar trouble in preceding generations.

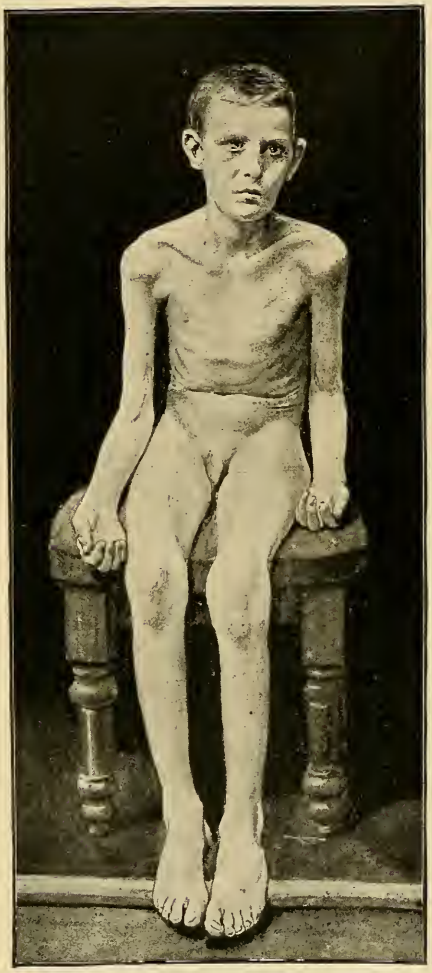


FIG. 78.—Case of Hereditary Ataxy (Friedreich's Disease).

The marked tendency to the occurrence of this disease in families is shown, as Gowers states, by the occurrence of sixty-five cases in nineteen families, giving an average of rather more than three to each fam-

ily. The number has varied from two to as many as eight in a single family. Both sexes are about equally liable, but sometimes a special predilection is shown in favor of one or the other sex. In one family of nineteen, two males were affected with the disease and seventeen females escaped. All the cases which I have observed were in boys, the sisters of these escaping entirely unharmed. The disease comes on, as a rule, very early in life, most of the symptoms being fully developed before the age of fourteen. In the family referred to by Everett Smith, of Boston, the symptoms of ataxic paraplegia were observed in the father at the age of sixty-six, but it is questionable whether his was a typical hereditary ataxy, and it is safer to adhere to the belief that Friedreich's disease will always appear early in life. If several members of one family are affected the disease is developed in all at about the same age.

FIG. 79.—Same Patient as in Fig. 78. Marked atrophy of the muscles about the shoulder girdles (infra-spinati, supra-spinati, rhomboids, and others).

SYMPTOMS.—The symptomatology of hereditary ataxy is now as firmly established as that of almost any spinal disease. Friedreich described most of the symptoms in 1861. The French school, including Charcot, Vulpian, Brissaud, and Marie, have given careful study to this disease. In England, Carpenter, Gowers, Ormerod, and Bury described cases of true hereditary ataxy; while in this country the chief contributions to this subject have been made by Smith, Hammond, Seguin, Dana, Church, Sanger Brown, and Inglis.

At a very early age children suffering from Friedreich's disease exhibit peculiarities in walking and standing. The child walks with its legs widely apart, in an uncertain, hesitating fashion, reminding one very strongly of a combined ataxic and cerebellar gait. There are in addition in some patients oscillatory movements of the head, which remind one a little of multiple sclerosis, and a little of such movements as we sometimes see in cases of senile degeneration. When the patient is asked to stand still, with his feet closely approximated, he soon begins to stagger, as tabic patients do, and as soon as his eyes are closed falls to the ground unless properly supported. Marie states that Romberg's symptom is ordinarily absent, but it has been most distinctly present in those subjects whom I have had occasion to examine. In addition to these disturbances in gait and in standing we also find a coarse tremor present in many of the cases, and in some a condition which is not so distinctly tremulous as it is ataxic and awkward. If the patient is asked to take hold of a pencil, to raise a glass of water to his lips, or to attempt to write, he seizes upon the object that he is to hold in a distinctly ataxic way and uses the hands awkwardly, sometimes after the fashion of extremely choreic movements. It is this combination of tremor, of awkward movements, and of choreic manifestations that gives to these children their characteristic aspect. There has been some discussion as to the occurrence of actual paralysis; in those whom I have examined there was in the first few years only a very slight loss of muscular power both in the upper and in the lower extremities. In advanced stages the grasp is almost nil, and the lower extremities may be so thoroughly paralyzed that it is impossible for the patient to lift a leg or even to move a toe, but in this respect the disease evidently varies very much.

Sensation may be interfered with to a slight degree, as is maintained by Senator, Oppenheim, Rutimeyer, Soca, and others. Lightning pains are infrequent, although Charcot insists that in some instances these occur quite as typically as in tabes. Anæsthesia and analgesia are rarely present. Several French authors have described cases in which hemianæsthesia occurred; but Marie is inclined to attribute this

to the association of hysteria with Friedreich's disease. The muscular sense is rarely affected, and in the persons whom I have examined for the purpose of determining the presence or absence of this sense, I have found it invariably present, even in those in whom there was marked incoordination of the upper extremities.

REFLEXES.—The superficial reflexes are, as a rule, preserved; but the deep reflexes are generally absent. The Babinski reflex is often present. The absence of the knee-jerks brings out in some subjects the very closest resemblance between Friedreich's disease and tabes dorsalis. In cases in which the reflexes are exaggerated there is good reason to doubt whether the disease corresponds to that described by Friedreich;* but there is no reason why in those forms in which the affection involves both the posterior and lateral columns the knee-jerk should not occasionally be augmented, and it is probably in view of such mixed types that Gowers has spoken of a form of hereditary ataxic paraplegia under the heading of Friedreich's disease. It is one thing to maintain a type as originally described by its discoverer, and quite another thing to deny that any departure from such a typical series of symptoms implies an entirely different order of disease.

Difficulties of micturition and of defecation occasionally occur, but are not so constant an accompaniment of Friedreich's disease as they are of tabes. Profuse salivation and disturbances in respiration (Oppenheim) have been recorded.

Trophic and vasomotor disturbances are also rare, if we except a general discoloration of the skin and coldness of feet and hands. Marie and other French authors refer to the occurrence of a special form of club-foot which they have observed in these cases of hereditary ataxy. Marie states that the foot is shortened; that the anterior portion is particularly broad if viewed laterally; the foot is in the condition of a pes cavus. The toes are hyperextended and have a claw shape. This deformity of the toes is said to have been observed as one of the early symptoms by parents in whose families this special disease has been hereditary.

* Some of these may belong to the Heredo-ataxie cerebelleuse of Marie.

Muscular atrophy constitutes an important symptom of this disease. It is most distinctly visible in the shoulder and pelvic girdles. It was so prominent a feature in one of my patients that on first examination I was inclined to regard the trouble as one of an hereditary form of progressive muscular atrophy, until the examination of the brother, who was in a more advanced stage of the disease, proved to me beyond the possibility of a doubt that both cases were typical of Friedreich's disease. In the boy represented in Fig. 90, the excessive atrophy must be attributed to an additional involvement of the gray matter of the cord.

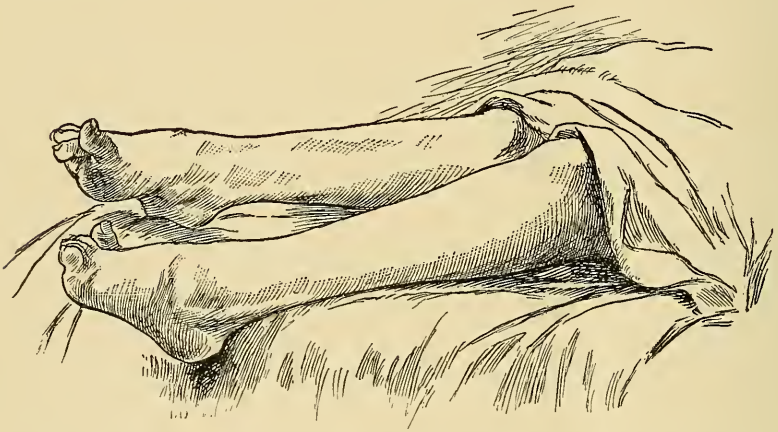


FIG. 80.—Deformity of the Feet in a Case of Friedreich's Disease; Hyperextension of the Toes and Club Foot. (Marie.)

We have now to consider a further set of symptoms which seem to me to prove the cerebro-spinal character of the disease. Nystagmus occurs in many of these cases, but, as a rule, does not appear until several years after the onset of the first symptoms. The nystagmus can often be elicited only upon extreme use of the ocular muscles. In this connection it may be well to remember that nystagmus upon extreme movements occurs in not a few healthy individuals, and that the presence of such movements in a child of a family affected with Friedreich's disease need not necessarily indicate the development of this serious trouble. Ocular palsies occur, but are rare. The Argyll-Robertson

pupil is as regularly absent in Friedreich's disease as it is present in cases of tabes. Optic atrophy and amblyopia have not been described in these cases. All the other senses remain entirely unaffected.

Among cerebral disorders occurring in the course of Friedreich's disease, vertigo and dizziness are extremely frequent. These may in part be due to the oscillatory movements of the head. In the earlier years of the disease the intelligence is not affected, except that the children cannot be educated at schools as other children are, and therefore remain backward. As the disease progresses a distinct defect of intelligence is noticeable, and a condition of semi-idiocy may be the result. The patients appear, however, far more idiotic than they really are, and this appearance is caused by the peculiar stupid expression of face, and above all by the disturbances in speech, to which we shall now refer. The speech reminds one a little of multiple sclerosis in that it is slightly scanning, but it is at times hesitating and jerky, at other times slow, deliberate, and awkward. On the whole, I find Marie's simile a good one, who compares it with the cerebellar gait, since it is, as he says, awkward, uncertain, and vacillating. All these symptoms are not developed until after the lapse of years. At first the peculiar gait and position are noticeable, later on the inco-ordination of the upper extremities and the difficulties of speech become more marked; then, after the lapse of a longer or shorter period of time, the patients become helpless and may remain bedridden or confined to their chairs for a period of ten and even twenty years.

DIFFERENTIAL DIAGNOSIS.—Attention must be directed to the following points:

TABES.	FRIEDREICH'S DISEASE.
Argyll-Robertson pupil present.	Argyll-Robertson pupil absent.
No nystagmus.	Nystagmus present in later years.
No tremor of the head.	Tremor of the head present.
No peculiarities of speech.	Speech altered.
Inco-ordination of upper extremities rare.	Inco-ordination the rule.
Lightning pains and various crises.	No such pains and crises, as a rule.
Ataxic gait.	Ataxic-cerebellar gait.
No defect of intelligence.	Intelligence defective in later years.

In the above parallel columns the symptoms which both have in common have been omitted, such as absence of reflexes and Romberg's symptom.

Multiple sclerosis can be distinguished easily from Friedreich's disease by the excessive exaggeration of the reflexes, by the intention tremor, by the marked spasticity of the gait, by the occurrence of ocular palsies, all of which rarely occur in cases of Friedreich's disease. Confusion might arise from consideration of the similarity in speech and the occurrence of nystagmus. Marie includes in the consideration of the differential diagnosis ordinary chorea, but it seems to me that no one but a very obtuse person could be misled by the occasional occurrence of awkward and choreic movements in Friedreich's disease, for in no other respect are the two diseases at all similar.

Confusion may more readily occur with cerebellar diseases, and Schultze some time ago, in answer to Senator's observations, called attention to the fact that, after all, cases that would seem to be of the cerebellar order are altogether different from the type which Friedreich described. I can approve of this remark of Schultze, for I have myself seen a case of defective cerebellar development* in which there was a superficial resemblance between it and cases of Friedreich's disease, and yet a careful consideration of the conditions was sufficient to distinguish between the two sets of diseases.

Pathological Anatomy. — The morbid anatomy of Friedreich's disease has not, as yet, been satisfactorily determined. Many autopsies have been made, but in some of the cases which have been examined post mortem, the symptoms have varied so much from the type, as laid down by Friedreich, that the results of the examination have helped to obscure rather than to clear up the doubts regarding the true anatomical changes of the disease. A number of authors have called attention to the attenuation of the entire cord in cases of hereditary ataxia, the diameter of the cord being but three-fourths or two-thirds of the normal. This attenuation is most marked in the dorsal region; but whether it is due to a disappearance of nerve-fibres, to the retraction of sclerotic tissue, or whether it is simply a defect of development, has not been clearly made out. Any

* See Chapter on Conditions due to Defective Development of the Brain.

one of these three causes may be operative in some cases, or a co-operation of all three is a possibility in others. The one fact that is absolutely indisputable is that on microscopical examination a sclerosis of the spinal cord is found, involving at different levels, or at one and the same level, various systems of the cord. This sclerosis affects most frequently the posterior columns or the lateral columns, or both together, and hence the symptoms vary between those of a pure posterior spinal sclerosis and those due to a

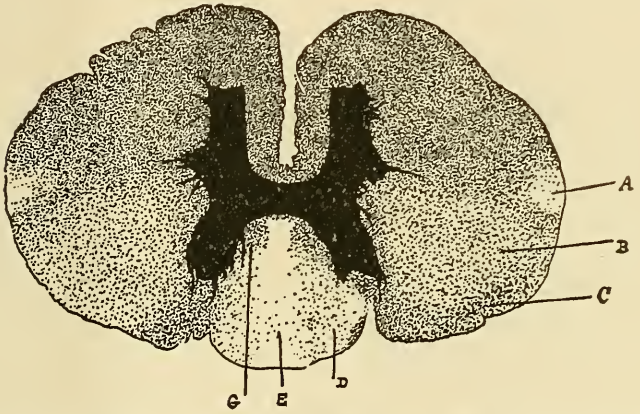


FIG. 81.—Section through Middle Dorsal Region of a Case of Friedreich's Disease. (After Blocq et Marinesco, from Marie's treatise.) A, small degenerated area, probably Gowers' tract; B, degeneration in lateral columns; C, direct cerebellar tract; D, column of Burdach; E, column of Goll — both degenerated; G, a strand of healthy fibres.

postero-lateral sclerosis, resembling the symptoms of the ataxic paraplegia of the adult.

Taking up the lesions of Friedreich's disease in detail, we may note the following distribution:

1. In the posterior columns, in a number of the cases, the columns of Goll are sclerosed throughout their entire length from the lower part of the spinal cord to the calamus scriptorius. The column of Burdach is also involved throughout the greater part of its course; but, according to Marie, the intensity of this sclerosis varies in different levels of the cord. The external portion of the column of Burdach, near the posterior horns, is generally exempt from sclerosis; in the cervical region the sclerosis of the column of Burdach begins to diminish, and disappears altogether in the vicinity of the medulla oblongata.

2. The direct cerebellar tract is involved from its beginning in the lower dorsal region to the upper cervical region. Toward this latter part it diminishes considerably, but is most marked in the upper dorsal segments. Several observers have noted that the lesion extends beyond the direct cerebellar tract, and involves the antero-lateral tract, or tract of Gowers; and Marie goes to the extent of stating that he thinks this involvement of the antero-lateral tract an almost constant feature of Friedreich's disease.

3. As for the lateral columns, the opinions of authors differ somewhat. Marie is not willing to allow the regular involvement of the lateral columns, while he concedes that the diseased fibres occupy the position of the crossed pyramidal tract. He does not believe that they represent the fibres of that

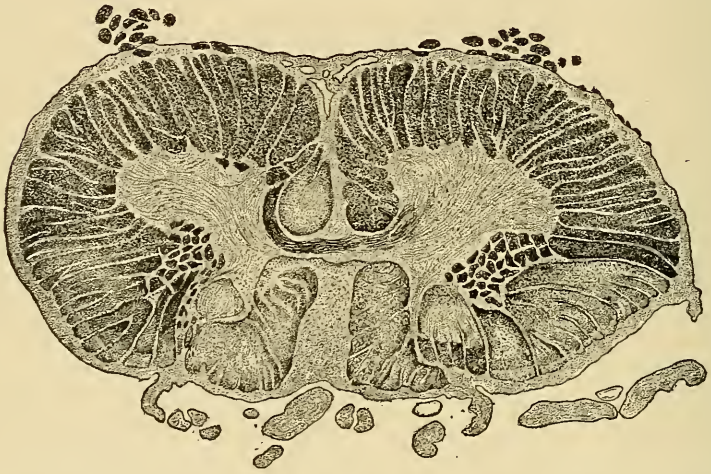


FIG. 82.—Section through a Cervical Segment from a Case of Friedreich's Disease. (Schultze.) Degenerated areas in posterior and lateral columns and in anterior column (left half of figure). Posterior root fibres also degenerated.

tract. His reasons for maintaining this are, first, that the lesion of the lateral columns in Friedreich's disease diminishes considerably from below upward to the level of the lower portion of the medulla oblongata, the very reverse of which would occur if these fibres were part of the pyramidal tract. Secondly, that on a transverse section of the spinal cord the localization of the lesion of the lateral column does not correspond exactly to the site of the lesion of the pyramidal tract proper. Third, nothing in the clinical appearances of Friedreich's disease reminds one of the symptoms which are a constant accompaniment of changes in the pyramidal tract. The only explanation which Marie is able to give for the fibres that are affected in the lateral region is that they are probably fibres which connect the direct cerebellar tract with the antero-lateral tract of Gowers. Of the reasons advanced by Marie, the first seems to me to be the only one that will bear close scrutiny; the second is scarcely susceptible of proof, and as for the third, it is sufficient to state

that the disease attacks both the posterior columns and the pyramidal fibres at various levels.

4. A difference of opinion also exists as regards the marginal zone of Lissauer, as many authors claim to have found these zones entirely intact, while others claim that an affection of these fibres constitutes a regular feature of Friedreich's disease. At all events the affection of Lissauer's tract is not nearly so constant or so early a feature of the disease as it is in the spinal ataxia in the adult.

The morbid anatomy of Friedreich's disease is not exhausted with this statement regarding the behavior of the white fibres. The gray matter is affected as well. The columns of Clarke exhibit a loss of fibres very much like that in tabes, as well as a considerable diminution of their cells, which are also smaller and devoid of their long-cell processes. In the posterior horns there is a diminution of volume, and also a decided diminution in the number of cells. The anterior horns have been found altered by some, Friedreich being among them, and the cells may be atrophied. There is no reason to doubt this occurrence in view of a considerable atrophy of the muscles which occurs in some of these cases, as in one of my own.

Some changes have been noticed in the ependyma of the central canal and in the cerebellum (Barker); yet these are not regular morbid features of Friedreich's disease. Some have also insisted that the spinal meninges are involved, while others dispute such involvement. If they are affected at all it is in the vicinity of the posterior columns. Little more can be said of the behavior of the posterior roots, although Blocq and Marinesco insist that they are as frequently altered in Friedreich's disease as they are in ordinary tabes. The peripheral nerves are supposed to be entirely normal in Friedreich's disease, and to this fact Dejerine and others attributed the entire absence of fulgurating pains in hereditary ataxy, while they are universally present in tabes dorsalis.

Senator laid especial stress on the congenital atrophy of the cerebellum and considered it to be of greater importance than all the spinal cord changes.

The question remains as to the nature of the morbid process underlying Friedreich's disease? Dejerine and Letulle, on the strength of certain peculiar fibres which they have found, and which they consider similar to fibres discovered by them in the cortex of epileptic patients, are inclined to regard the sclerosis of hereditary ataxia as a sclerosis of the neuroglia. They insist that neither the connective-tissue septa nor the blood-vessels are at all altered, and that the sclerosis is therefore very different from the vascular sclerosis which so often involves the pyramidal tracts and the direct cerebellar tract; but later investigators, among them Weigert, have disproved these views of Dejerine and Letulle.

They regard the sclerosis of Friedreich's disease as in no-wise different from the ordinary sclerosis excepting in this, that it has been developed at an unusually early period. Weigert, in his recent studies on the neuroglia, concludes that in the ordinary spinal sclerosis, as it occurs in tabes, in multiple cerebro-spinal sclerosis, and in amyotrophic lateral sclerosis, the proliferation of the neuroglia is a much more marked feature than in hereditary ataxy. The refutation of this theory of Dejerine and Letulle does not, however, help us in the recognition of any true theory of the disease, and for the present we must confess that we do not know why or how the sclerosis of Friedreich's disease is developed.

PROGNOSIS.—The prognosis of Friedreich's disease is invariably grave. In the course of four to six years the patient becomes crippled, unable to move about except in a chair, awkward in speech, and more or less demented. Unfortunately the disease does not lead to a fatal issue within a reasonable period of time, some of the patients attaining the age of forty years and more.

As for treatment, no special method need be recommended. The progress of the disease cannot be stayed, the relief of pain constituting the only reason for active interference on the part of the physician. The ordinary anti-neuralgic remedies may be employed to this end. In addition, suspension, electrical treatment, massage, and the like, may be ordered, in the discretion of the physician.

HEREDITARY ATAXY (CEREBELLAR TYPE; TYPE NONNE–MARIE).

Under the title "Heredo-ataxie Cerebelleuse" Marie collected a number of cases which bear a very close resemblance to Friedreich's disease, yet present several characteristic and different symptoms. Titubation, Romberg's symptom, tremor of the head and of the extremities, choreiform, awkward movements, particularly of the arms and hands, and nystagmus, are symptoms common to Friedreich's disease, and to the form we are now describing. The cerebellar form of hereditary ataxy comes on, as a rule, at the age of puberty or later. The knee-jerks, instead of being diminished, are increased. There are marked ocular symptoms, the pupils fail to react to light and during accommodation. There is diplopia in some cases, color-blindness in others. Contraction of the visual field has been observed, and amblyopia, due to atrophy of the optic nerve, is a typical symptom. There are also marked

disturbances of sensation. The deformities of the foot and kyphosis, noted in Friedreich's disease, are never present in the cerebellar type.

This symptomatology, as developed by Marie, reveals a very striking resemblance between these cases of hereditary cerebellar ataxy and a form of family disease described by Nonne. In all probability Nonne's and Marie's descriptions refer to the same type. While the latter deserves credit for naming the disease, it seems to me just that Nonne's name should be connected with this type. There is also a very close resemblance between this hereditary cerebellar ataxy and some of the cases which have been described under the heading of "Hereditary Spastic Paralysis." Senator's views regarding Friedreich's disease may in part be explained by the recognition of this special order of disease, and there would surely be a possibility of confounding this type of hereditary ataxy with the cerebellar type of multiple sclerosis which Charcot has described.

It is too early to report definitely upon the pathology of hereditary cerebellar ataxy. If we are correct in considering the cases described by Marie and Nonne as representatives of the same type, the post-mortem findings in one of Nonne's cases will give some clew to the pathology of this variety of disease. In Nonne's case the cerebellum was small, and there was a simple diminution of tissue without sclerosis or degeneration. The spinal cord was much attenuated throughout its entire length. Under the heading of "Hereditary Ataxy and Atrophy of the Cerebellum" Menzel described a condition in which there was a combined systemic disease of the spinal cord, and in addition atrophy of the cerebellum, pons, and medulla. Two French authors, Royet and Collet, have reported a case which was diagnosticated during life as multiple sclerosis, in which there was found on post-mortem examination a lesion of the cerebellum and of the cerebellar tracts in the pons. It is possible that some of the cases which have been regarded as a form of hereditary multiple sclerosis may come under this same heading. I recognize that there is great danger in multiplying types of disease. The form described by Marie and Nonne should in justice to these two authors be given due consideration; but I believe it better for the present to regard it as a variety of hereditary ataxy.*

HEREDITARY SPASTIC PARALYSIS (SPINAL, CEREBRAL OR CEREBRO-SPINAL TYPES).

During the past decade or two several family affections have been described by Homén, Newmark, Pelizaeus, Strümpell, Freud, Erb, and myself, in all of which a spastic rigidity, or a spastic paralysis affecting the lower extremities chiefly, has been the most prominent symptom. In some cases the spastic symptoms were evidently of purely

* The cases reported by Dr. Sanger Brown also resemble this type.

spinal origin; in other cases they were associated with distinct cerebral symptoms. A classification of all of these various diseases according to the morbid conditions underlying them, is not yet possible; it is sufficient for the present to discuss them from the etiological point of view. I have chosen, therefore, to designate them as *hereditary* spastic paralysis, and to subdivide them, according to their more prominent symptoms, into a spinal, and a cerebral or a cerebro-spinal variety.

After adopting a subdivision of this kind, the pathology of each type will be more readily made out; and it is quite probable that the various types will be found to represent a mere difference in the topographical distribution of lesions and symptoms, while the morbid process, an arrest of development, or an early degeneration, is common to all.

The spinal type of hereditary spastic paralysis is well illustrated by Newmark's cases. In view of the importance of the subject a short summary of a few of his cases will be given.

A girl, aged fifteen, tall and intelligent; normal birth and labor; no convulsions. Did not kick with the legs as other children do in bed. Began to walk at eighteen months. Gait peculiar from the first, and always remained typical of spastic paraplegia. Lower extremities flexed at the hips and knees; adduction of thighs and pes equinus. No wasting of muscles, which react well to both currents. Knee-jerks and adductor reflex exaggerated; no ankle clonus (probably on account of extreme contracture). No disturbance in the upper extremities, but all tendon reflexes exaggerated in them. Jaw-jerk present. Intelligence, sensations, and sphincters intact. No nystagmus, no strabismus. Girl is able to use a tricycle. Speech is normal. A brother, aged five, exhibits very similar symptoms. No history of syphilis in the family. There were no other cases of similar nature in the family, except one of spastic diplegia in a first cousin; but this diplegia was evidently the result of difficult labor.

In the second family the father is said to be a tall, healthy man of thirty-eight, who denies syphilitic infection or alcoholic habit. Has never known of any affection similar to that observed among his children. His reflexes are normal. Mother of children, aged thirty-seven, has always been healthy. Her knee-jerks and Achilles tendon reflex quite active. Lively reflexes in the upper extremities and jaw-jerk distinct; no ankle clonus. No blood relationship between man and wife. They have had eleven children, eight of whom are living. The oldest living child is a boy, aged sixteen, who was well until about a year and a half before he was examined. At that

time a stiffness appeared in the legs, especially on rising in the morning. His condition gradually developed into a mild form of spastic paraplegia, with typical gait and position of the extremities. Knee-jerks much exaggerated; no patellar clonus nor ankle clonus. Plantar, cremasteric, and abdominal reflexes very lively. Abdominal muscles said to be rigid. Upper limbs normal; reflexes much increased. Jaw-jerk well marked. In a second brother, aged fourteen, the symptoms had become more pronounced, and had set in at the age of seven and a half years. The legs were stiff and walking was difficult; had been compelled to use crutches. There were contractures in the knee-joints; there was adductor spasm and pes equinus, furthermore, an exaggeration of the knee-jerks. Ankle clonus was present, and the plantar and cremasteric reflexes were also very lively. A third brother developed exactly the same symptoms at the age of nine years, after typho-malarial fever. Though these three cases are the most pronounced ones in the family, one sister exhibits increased reflexes; a boy, eight years of age, is said to be stiff in the knees, with exaggeration of the patellar reflex; another child, aged six, is said to have a lazy, dragging walk, with excessive increase of knee-jerks; a child, aged three and one half, has active knee-jerk and jaw-jerk.

That there is a tendency to spastic paralysis in this family there is no doubt; but the fact that instruments were used at the birth of several children, including the one whose symptoms were most pronounced, makes it probable that "heredity" alone was not responsible for the multiplicity of cases. In a more recent publication (1904) Newmark reports upon the development of the children referred to in his earlier articles.

The symptoms of this spinal type (spastic paraplegia, with rigidity or contractures, increased reflexes) are distinctly due to interference with the pyramidal tracts in the lateral columns of the cord. Many years ago Strümpell described the cases of two brothers, who presented the typical features of a spastic spinal palsy. In one of these cases a post-mortem examination was made, by which it was determined that the symptoms were due to a primary systematic degeneration of both pyramidal tracts in the lateral columns, together with a slight affection of the cerebellar tracts and the columns of Goll. More recently Strümpell has described another case, very much like the other two, in which he made the diagnosis of hereditary spastic palsy. In the family of this patient the grandfather was said to have suffered from a palsy of the

legs, the father had a peculiar gait, and people who knew both the father and the present patient said that one walked like the other. Two uncles were said to have the same walk. The mother of the patient and other brothers and sisters were entirely healthy. The patient did military service from his twenty-first to his thirty-third year, and was not inconvenienced in any way, except that he occasionally noticed a peculiar feeling in the legs. At the age of about thirty-five his first symptoms began. His legs became stiff and he soon had to resort to a stick in walking. The examination of the patient revealed a typical spinal spastic paralysis, with exaggeration of the reflexes, rigidity of the joints, normal sensation, and a spastic gait. The vesical and rectal reflexes were not interfered with. All the symptoms steadily increased, but no new ones were developed. In addition to its interest as a form of hereditary spinal disease the type also deserves some special recognition from the fact that, from first to last, the symptoms have been such as are due exclusively to disease of the lateral columns. Strümpell refers to Bernhardt's patients, and claims that they are entirely identical with those described by him. Four out of six brothers presented the phenomena of spastic spinal palsy; three of these four developed their symptoms at the age of thirty, and all the morbid signs progressed in a remarkably slow fashion. I can agree with Strümpell in believing that these diseases are entirely similar to those described by him, although in one case of Bernhardt the symptoms tended toward a cerebro-spinal rather than a purely spinal type. The forms described by Tooth and Philip should be placed in the same category.

Spastic spinal paralysis (spastic paraplegia) occurs in children of families in which there is no history of a similar affection in the same or past generations. In February, 1893, a girl, four years of age, was brought to my clinic, who was afflicted with a typical spastic paraplegia; paresis and rigidity of both lower extremities; very rigid gait, increased knee-jerks on both sides, slight double ankle clonus—these reflexes being a little more marked on the right side than on the left side; increased reflexes in the upper extremities, but no weakness. Electrical reaction of all muscles and nerves normal; no disturbances of sensation; no impairment of vesical and rectal reflexes; no nystagmus. Intelligence good. The birth of the child (first-born) had been entirely normal and at full term; no instruments were used; the child was

supposed to be perfectly well, except that it did not begin to walk until it was three years old, and its walk was unusually stiff and awkward; it dragged its feet. Three younger children are perfectly healthy. Excepting the early age of onset there is no distinction in clinical symptoms between such cases as these and the spastic spinal paralysis of the adult.

MORBID ANATOMY.—In one of Newmark's cases there was a distinct degeneration of the pyramidal tracts, most marked in the lumbar portion of the cord; a degeneration of the posterior columns, most marked in the upper dorsal region; also an involvement of the columns of Clarke—in short, a combined degeneration not unlike the findings in Strümpell's cases and in Friedreich's ataxia. In view of Strümpell's investigations there is no longer any doubt about the occurrence of a primary degeneration of the lateral columns developing later in life, but due in all probability to abnormal congenital conditions. There is some reason, therefore, to believe that the question of the occurrence of a primary lateral sclerosis will be affirmatively answered through these studies on hereditary spastic paralysis.

DIAGNOSIS.—The diagnosis of these diseases need not be given in further detail. The history of a case exhibiting the symptoms of spastic paraplegia of the lower extremities, with increase of the reflexes, with rigidities and contractures, without involvement of the vesical or rectal reflexes, without atrophy, without disturbances of speech and nystagmus, would be sufficient to place the case in this category, provided the hereditary character of the affection could be established. The disease should be carefully differentiated from the spastic cerebral palsies of childhood.* The diplegias and paraplegias occurring as a result of diffi-

* In an article on Hereditary Spastic Paralysis, Erb stated that some of the congenital diplegias and paraplegias which Freud, myself, and others have attributed to cerebral lesions may at times be due to spinal lesions. I am willing to concede the possibility of this; but I think that such spinal affections are, after all, rare; and there is the one fact, to which I have alluded in various writings, that in forty-five per cent. of all the cases of infantile spastic paraplegia there is marked idiocy. This association militates against the diagnosis of a purely spinal disease. I have reviewed the histories of all my patients with supposed cerebral paraplegia and can find none which I am willing to attribute to a spinal lesion. The child reported on page 348, though suffering from paraplegia, was recognized as probably of spinal origin; and such an origin may be suspected in types of spastic paraplegia, without idiocy coming on several years after a normal birth.

culty during labor (Little's disease), would be most easily confounded with this hereditary form. A careful inquiry into the history of labor, the determination of the exact age at which the trouble began, the frequent occurrence of convulsions and of defective mental development, in birth palsies, will help to differentiate between these two sets of cases. If spastic paraplegia is developed in a child, or in a youth, that has not been preceded by convulsions, or is not associated with defective mental development, the case would come more properly under the head of hereditary spastic paraplegia, and we must remember that such cases may occur without any history of similar disease in the same or preceding generations.*

The prognosis of hereditary spastic spinal paralysis is a grave one as regards recovery from the disease, but not unfavorable as regards the duration of life, for the majority of patients thus far examined have been well advanced in years. In the way of treatment, nothing can be attempted except to apply the usual methods of massage and electricity, or possibly to attempt to improve contractured limbs by various tenotomies and by transplantation of tendons.

CONGENITAL SPASTIC PARAPLEGIA. (LITTLE'S DISEASE.)

In this connection brief reference must be made to an affection which is generally spoken of as "Little's Disease." It refers to a condition of spastic rigidity of the limbs with paralysis which is observed in the earliest months and years of life, and is almost invariably due either to defective development or to injuries received during protracted labor. Van Gehuchten, without sufficient justification, restricts the term to the disease occurring in children born before full term. As the present author showed in 1897, so many different conditions have been collated under the term *Little's Disease* that much confusion has resulted. It will be well, therefore, to quote from Little's original paper in "The Transactions of the London Obstetrical

* Marie and some other French authors deny the existence of a primary spinal spastic paralysis in the adult, and believe that if such a condition does exist, it is to be traced back to the early years of life. They describe as "Tabes dorsal spasmodique" of children, a group of symptoms which German and American authors have included under the term, Congenital Spastic Diplegia and Paraplegia. The French view would take all of these cases out of the category of cerebral diseases, and for this there is as yet no warrant. Marie (p. 101) states that "Tabes dorsal spasmodique" is never hereditary; and attributes the condition to defective development of the pyramidal tract. This tract is surely a cerebro-spinal affair.

Society," Vol. 3, 1862, in which he says: "I showed that premature birth, difficult labors, mechanical injuries during parturition to head and neck where life has been saved, convulsions following the act of birth, were apt to be succeeded by a determinate affection of the limbs of the child which I designated, spastic rigidity of the limbs of new-born children, spastic rigidity from asphyxia neonatorum and assimilated it to the trismus nascentium and the universal spastic rigidity sometimes produced at later periods of existence."

He speaks of spastic rigidity being sometimes associated with paralytic contraction. He evidently had a paralytic condition in mind, and in that respect various recent writers who seem to deny the existence of paralysis in these cases and wish to speak of them only as spastic rigidities, are in the wrong.

In association with Dr. Peterson I showed many years ago that the frequent occurrence of mental defect with congenital spastic rigidity and paralysis was due to the various cerebral lesions and accidents which are responsible for other cerebral palsies of children. The symptoms are, as a rule, limited to the lower extremities, although on examination some slight difficulty in the upper extremities may frequently be noted. Cross-legged position and cross-legged gait are typical symptoms of this trouble. There is, of course, close resemblance to disease involving the lateral columns of the cord, and in this respect the cases must be carefully differentiated from primary or hereditary spastic paralysis and from syphilitic spinal paralysis.

Recent observations of Dejerine and of Spiller have shown that congenital spastic paraplegia may be due to changes in the lateral columns of the cord only, without any cerebral disease, but in spite of these two trustworthy findings, the present writer still believes that in most instances the conditions which could reasonably be designated as Little's Disease, are to be attributed to defective development of, or to lesions in, the brain. Further information on this subject can be gathered from the chapter on infantile cerebral palsies. It will be recognized that if the term Little's Disease is to be retained at all, it should be applied only to the cases of spastic paraplegia, or of spastic diplegia (complete or incomplete), due to premature birth or to difficulties during parturition.

CHAPTER XXI.

PROGRESSIVE MUSCULAR ATROPHIES.

In this chapter we intend to discuss all those diseases which are characterized by a progressive weakness and atrophy of certain groups of muscles. We have nothing to do with muscular atrophy, whether progressive or not, which follows after *acute* disease of the brain, of the spinal cord, or of the peripheral nerves. The term "progressive muscular atrophy" was formerly given to a single type of disease with which we shall become more intimately acquainted, and for a time all cases resembling this one type were designated in the same way. It became evident, however, that this one term was altogether too general. While it was a convenient clinical designation for an entire group of diseases, it did not sufficiently describe a number of other forms which were closely allied to the chief type. In order to avoid confusion, it would be well if we could dismiss the term progressive muscular atrophy altogether, for in many of the cases hypertrophy as well as atrophy is present for a long period of time. The word dystrophy, which might be used to designate both conditions, has, unfortunately, been restricted to the cases of primary or idiopathic muscular wasting; we cannot, therefore, apply it to the spinal forms. Moreover, a number of diseases were included under the heading of progressive muscular atrophy which we now recognize to have been cases of amyotrophic lateral sclerosis, of syringomyelia, and possibly of spinal syphilis.

The chief question at the present time regarding the various forms of progressive muscular atrophy is, whether in a given case the disease is of spinal or muscular origin.

Those forms of progressive muscular atrophy due to a spinal lesion we call *amyotrophies*, and to those due to disease of the muscular system alone, we give the name *myopathies*. We shall see that there is some reason, too, to constitute a third type, which we might designate as a neural form of progressive muscular atrophy. (Fig. 83.)

The study of this entire subject began many years ago with the establishment of two distinct diseases—the first was the typical "progressive muscular atrophy," as described by Aran and Duchenne; the second, pseudo-hypertrophic muscular paralysis. Since that time at least six different forms of progressive muscular wasting have been described, and the attempt has been made in each case to prove the relation of the special form, either to the Aran-Duchenne type, or to the type of muscular pseudo-hypertrophy. The Aran-Duchenne type has become the chief exponent of progressive amyotrophies; while muscular pseudo-hypertrophy has been considered the most pronounced form of primary myopathies. The various types of progressive muscular disease have been established very largely in accordance with the mere topographical distribution of atrophy or hypertrophy. Though convenient for clinical designation, such a distinction is not sufficient for a rational classification of these various diseases. It must be our aim to find the cardinal symptoms which will help us to differentiate at once, and easily, between those cases of progressive muscular disease due to spinal-cord lesions and those primary dystrophies, which represent a disease of the muscular system.

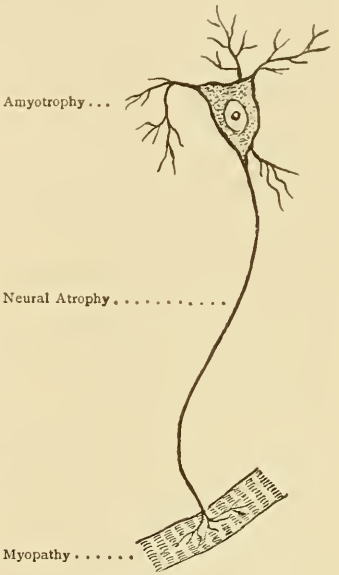


FIG. 83.—A Diagram designed to show the Site of the Morbid Lesion in the Several Groups of Progressive Muscular Atrophy.

The following are the cardinal symptoms present in the majority of cases belonging to the two large groups of cases :

Progressive Amyotrophies.	Progressive Myopathies.
Onset late in life; rarely in early childhood.	Onset in early life.
Not hereditary, as a rule.	Generally hereditary (family trouble).
Wasting first in the upper extremities (leg type rare).	Wasting or hypertrophy begins in the lower extremities.
Hypertrophy does not occur.	Hypertrophy frequent.
Fibrillary twitchings.	No fibrillary twitchings.
Reaction of degeneration often present in affected muscles.	Reaction of degeneration rare (quantitative, not qualitative, electrical changes).

These points of differential diagnosis will hold good in the majority of cases. Werdnig, Hoffmann, and others have reported cases of spinal progressive muscular disease due to hereditary or family influences. Fibrillary twitching has been seen in some cases that appeared to be typical myopathies, and the electrical reactions have been found considerably altered in similar cases, so that of all these cardinal symptoms there are, after all, only a very few which are invariably present in one or the other form of progressive muscular disease, and it is wiser, therefore, to be guided by the general agreement of symptoms rather than by any one single symptom.* This confusion of types of disease and of symptoms, need not cause surprise, if we remember that the ganglion cells of the spinal cord, the peripheral nerves, and the muscles, constitute a physiological unit (Neuron).

With these prefatory remarks we may proceed to the consideration of that class of cases which for a very long time were supposed to be the only representatives of what was formerly called "progressive muscular atrophy." This is a spinal-cord affection which, as a rule, begins late in life. It might, therefore, be considered out of place to treat of this disease in a work on the nervous diseases of children; but the entire subject of muscular diseases cannot be properly understood unless we can recognize this special type, and, furthermore, cases of this type have of late years been described by Hoffmann and others, in children.

* The hereditary cases reported by Werdnig are particularly interesting in this respect.

PROGRESSIVE AMYOTROPHY.—"PROGRESSIVE MUSCULAR ATROPHY." (TYPE, ARAN-DUCHENNE.)

This form begins, in the majority of the cases, with an atrophy and a corresponding weakness in the small muscles of the hand (thenar and hypothenar). The atrophy extends slowly from muscle to muscle ("atrophie individuelle"), beginning as a rule with the adductor pollicis. It involves by degrees the opponens pollicis and the deep muscles of the thenar. From these it gradually extends to the muscles of the hypothenar, the interossei, the flexors and extensors in the forearm. At this point the disease may remain stationary, or it may spread to the flexors in the upper arm, to the deltoid, the triceps, and finally to the muscles of the trunk, the shoulders, and the back. Duchenne recognized the fact that the atrophy may, in exceptional cases, begin in the trunk, in the shoulders, or in the legs. Some of these would now, no doubt, be considered under a different heading, but I have myself seen cases which could in nowise be distinguished from the typical Aran-Duchenne disease which began in the muscles of the thighs. If the disease begins in the upper extremities, the legs are, as a rule, not affected until very late in the course of the disease; but there are exceptions to this rule. I have observed several cases in which the atrophy in the lower extremities began almost simultaneously with that in the upper extremities.

The atrophied muscles in this form exhibit fibrillary contractions, and, as a rule, present marked changes in electrical contractility. These changes are not so pronounced as in cases of acute or subacute anterior poliomyelitis, and yet after the disease has lasted for a considerable length of time a typical reaction of degeneration will be found present in most of the wasted muscles. In the majority of cases thus far recorded the disease has not been distinctly hereditary, although series of such cases with distinct hereditary tendencies have been published by Naunyn, Eichhorst, Hammond, Osler, and others.*

* The famous Weathersby family, reported by Hammond, and the Farr family, of Vermont, described by Osler, may possibly represent other types of progressive muscular atrophy, which we shall consider later on.

As the disease progresses the wasting becomes more and more extreme; the patient is no longer able to get about, becomes bedridden, and after many years of annoyance, if not of suffering, dies of some intercurrent disease, or from extension of the process upward into the region of the medulla, with consequent paralysis of the vital centres.

The symptoms of the disease were well described by Duchenne, and very little has been added since his day to the clinical characterization of this special form. But a very warm discussion was waged for a long time regarding the origin of the disease, some maintaining its spinal origin, others believing it to be a peripheral disease. There was some reason for this difference of opinion, for the cases upon which the older authors based their views were in part due to spinal disease, and in part due to nerve or muscular lesions. The microscopical studies of Charcot and Joffroy, of Lockhart Clarke, of Hayem, and others, proved beyond a doubt that this special form of progressive muscular atrophy was due to changes in the spinal cord. The chief changes found are these: A sclerotic and pigmentary atrophy of the ganglion cells of the anterior horns, inflammatory changes in the neuroglia, increased size of the blood-vessels, and proliferation of the cellular elements. In fresh preparations granular corpuscles are found, and according to the degree and stage of the disease the anterior horns may be very much reduced in all diameters, and the ganglion cells either atrophied or entirely lost. The anterior nerve-roots are affected secondarily to the lesion of the gray substance. The nerve-fibres are not all destroyed, a number of them remaining intact. Those that are destroyed exhibit the appearances of simple atrophy, a point to which Charcot alludes as distinguishing these cases from infantile spinal paralysis.

According to these pathological findings we must suppose that an inflammation spreads slowly from the ganglion cells of the anterior horns along the anterior nerve-roots without destroying as many of these fibres as is the case in infantile poliomyelitis. The atrophic changes in the muscles are the direct result of irritation, which begins in the cells of the anterior horns, and is propagated thence through

normal or only half-wasted nerve-roots, to the peripheral muscular fibre. Positive as these anatomical findings seem to be, it is somewhat surprising to learn how few reliable post-mortem examinations have been made in these cases proving the correctness of these views. The cases of Pierrot-Troissier, of Strümpell and of Hoffmann, are among the few which have been so carefully examined as to have placed the spinal origin of this special type of progressive muscular atrophy beyond question. In these cases the anterior gray matter was the only part affected, and alone responsible for the wide-spread muscular atrophy.

There is no need in this treatise to give a typical history of a case of spinal progressive muscular atrophy, as it occurs in the adult. I have stated above that these cases are, as a rule, not hereditary, and upon this absence of the factor of inheritance the differential diagnosis was formerly frequently based between a progressive amyotrophy and a progressive myopathy. But this point of differential diagnosis has been rudely shaken by the interesting articles which Hoffmann and Werdnig published some years ago concerning the occurrence of chronic spinal muscular atrophy in children. I will endeavor to summarize one of Hoffmann's cases of

HEREDITARY PROGRESSIVE MUSCULAR ATROPHY.

A girl, four years of age; the birth of the child was entirely normal; when nine months of age was able to stand; early abnormal development of adipose tissue. Gradually the child became so weak that it could not stand, could not sit upright in bed, could not turn around without assistance. For a long time it was able to move its feet and its arms. The motor disturbances increased gradually, and the child lost its superfluous fat and became thoroughly emaciated, particularly in the trunk and the extremities. The face remained full. The sphincters were at no time involved. Mental development was good. Speech was normal; no convulsions; no strabismus; no difficulties in mastication or deglutition. The child was able to turn its head, but could not lift it from the pillow. There was no evidence of any hypertrophy or pseudo-hypertrophy in any of the muscles, but there was paresis and atrophy of the deep muscles of the neck, of the sterno-cleido-mastoid, of the trapezius, of most of the shoulder-muscles, of the latissimus dorsi, of the serrati, the pectoral and deltoids, and of the flexor muscles in the arm. The biceps and brachialis anticus were very thin, the supinator longus a little stronger than these. The triceps was thin and weak. The extensors and

flexors in the forearm were also atrophic and paretic. The thenar and hypothenar eminences were thin, flaccid, and weak, as were also the individual interossei. The paralysis was in proportion to the atrophy of these muscles. The tendon reflexes were entirely wanting in the upper extremities. The mechanical excitability of the muscles was diminished. The nerve-trunks were neither thickened nor sensitive on pressure. The paresis and atrophy of both upper extremities were entirely symmetrical. There were no fibrillary movements. There were no trophic or vasomotor disturbances of the skin in the upper extremities. Sensation was entirely normal. There was marked diminution of electrical excitability in the median and ulnar nerves, and complete reaction of degeneration in the biceps.

The muscles of the back and abdomen were very paretic, the long muscles of the spine much diminished in volume and power. There was lordosis of the lumbar region of the spinal column. The gluteal muscles, and all the muscles of the thigh, were very atrophic and almost completely paralyzed. The muscles of the leg were also atrophic and paretic. Movements of the toes tolerably good. A progressive diminution in the volume of the calf and thigh muscles was noted at periods six months apart. No indication of any deep reflexes in the lower extremities. The faradic excitability of the nerves in the lower extremities was distinctly diminished. No sensory disturbances, no fibrillary or fascicular or choreic movements of muscles. The paralysis was symmetrical and flaccid. The joints were like those of infantile spinal palsy. The child died of an intercurrent pulmonary trouble about one year after the first examination.

The report of the autopsy included the following points: The lumbar portion of the spinal cord less in volume than under normal conditions. Very marked atrophy of the anterior spinal-cord roots throughout the entire spinal cord as high up as the medulla. On microscopical examination the chief changes found were in the anterior gray matter and in the anterior nerve-roots, from the lower portion of the medulla through the whole spinal cord. There was a distinct atrophy and diminution in number of the ganglion cells of the anterior horns throughout the entire spinal cord. This was more marked in the lumbar than in the cervical portion. There was also very marked atrophy of the anterior roots and a similar affection in the peripheral nerves and the nerve-filaments in the muscles, as well as a very marked atrophy of the muscles supplied by these nerves. In addition to these chief changes there were also slight and symmetrical changes in the motor tracts of the spinal cord, particularly in the crossed pyramidal tracts and in the lateral columns as well as in the direct pyramidal tracts. Medulla oblongata was not involved.

This case is so similar, from a clinical and anatomical point of view, to the typical cases of the Aran-Duchenne type that the close relationship between these forms cannot be doubted. It remains to add that an entirely similar affection was reported in the case of a brother of the first child, in whom the disease began at about the same age and behaved in very much the same way. The parents of these two children have raised a family of fifteen; several of these have died of convulsions, others are afflicted with

distinct lipomatosis, and the parents invariably accepted the occurrence of this excessive accumulation of fat as an evil omen.

Abortive and hereditary forms of progressive muscular atrophy of the spinal type have come under my notice. The cases are those of a physician, living in Canada, and his daughter, about twelve years of age; in both of them there was distinct atrophy of the interossei of both hands, flattening of the thenar, and slight extension of the atrophy to muscles of the forearm. "Weak hands" have been characteristic of the family, but the disease does not appear seriously to invade other parts. The father (physician) has had this trouble for years, and has been compelled to give up surgical work.

PROGRESSIVE NEURAL MUSCULAR ATROPHY—PROGRESSIVE NEUROTIC MUSCULAR ATROPHY—THE PERONEAL FORM, OR LEG TYPE, OF PROGRESSIVE MUSCULAR ATROPHY.*

In this form of progressive muscular wasting the disease begins, in the majority of cases, in the lower extremities. At first the extensor muscles of the toes show a slight weakness. The small muscles of the feet may become involved, and then the atrophy spreads very much after the fashion of the spreading of the atrophy in the spinal cases, from muscle to muscle, until the entire leg is considerably atrophied and weakened. As a result of this weakening, deformities of the foot may arise; pes equinus or pes equino-varus is a frequent result. In other cases a distinct club-foot is developed, and inasmuch as the affection may spread quite rapidly from one side to the other, a progressive form of wasting in both lower extremities, including possibly the development of double club-foot, is extremely suggestive of this "leg type" of progressive muscular atrophy. The disease, as Hoffmann has shown, in rare instances attacks the upper extremities first and then involves the lower. Hoffmann has objected to the use of the designation "leg type," but since in this form the legs are involved at a very early stage, whether the disease begins in them or in the upper extremities, it seems proper for the present to retain this designation. The atrophy in the upper extremities may involve the small muscles of the hand, the extensor and flexor muscles of the forearm or the

* This disease is also known as the type of Charcot-Marie-Tooth. Hoffmann suggested the term "progressive neurotic," Bernhardt, the term "progressive neural," muscular atrophy.

arm, and may cause a wasting of the muscles about the shoulder-girdle. I have seen the infra- and supra-spinati especially wasted in several of these cases. The atrophy in the upper extremities is, as a rule, not so distinct nor so early a symptom as in the cases of the Aran-Duchenne type.

FIG. 84.—Two Brothers afflicted with the Peroneal Form of Progressive Muscular Atrophy, Eight Months and One Year respectively after First Operation.

Sensory changes are generally present, and serve as an important point of differentiation between this special form of atrophy and a spinal amyotrophy. The various forms of sensation may be slightly altered, or, in some cases, tactile sensation and temperature sense may remain normal, while the pain sense may be more distinctly involved. Paræsthesiæ may be present in addition to the objective changes in sensation. The reflexes in the lower extremities are either diminished or lost; the exact state of the reflexes

depending somewhat upon the stage of the disease at the time the patient is examined. The electrical reactions in the atrophied muscles are, as a rule, altered. The changes are not so extreme as in the cases of spinal amyotrophy, nor are they as mild as in the primary muscular dystrophies. The reactions are diminished quantitatively, and altered also as regards the quality of contractions. A case has not yet been reported in which the muscles of the face were involved, and there were no changes in sensation, and none in electrical reactions in any of the muscles or nerves of the neck and head.

The symptomatology of this rare form will be best elucidated by an extract from the account given of the disease as it occurred in two brothers which was reported in a paper published by the author in the year 1890. These cases are all the more interesting as they are the only ones, to my knowledge, which for a time were successfully treated by surgical measures.

The family history is very meagre. The father, a Bavarian, is dead; cause of death unknown. Mother, living and healthy, thirty-two years of age. The two patients were the only children. Both boys were born healthy. Each showed disturbances in the use of the legs at a very early day, and at the age of five years both had acquired double club-foot.

When the younger brother was first admitted to the hospital, in 1887, he was compelled to use crutches. According to the hospital records there was marked shortening of the Achilles tendon and plantar fascia of both feet. Foot arched (pes cavus); when at rest inner side does not touch the floor.

Measurements: Right calf, 7 inches; left calf, 7 inches. Dr. Gibney performed double achillotomy. Separation of ends 1⅞ inch on the left and almost the same on the right side. Feet were flexed dorsally to about eighty degrees, and plaster-of-Paris splints were supplied. He was discharged four months after the operation, with a note that the patient walks quite well, soles flat on the ground, toes slightly inverted. He returned to the hospital in 1888 with paralytic limp and with a position of the feet as represented in the accompanying figure. Double achillotomy was again performed, division of plantar fasciæ was made, and there followed application of Thomas's tarsoclast and plaster splints with the results as shown in Figs. 101 and 102. Two months after the operation the feet were in typical calcaneus position, when using his shoes without apparatus; standing squarely on the soles of the feet he shows disposition to roll feet inward. He could voluntarily flex the ankle-joint a little beyond ninety degrees, but in so doing the toes were hyperextended. He walks very much as children do with a peroneal type of poliomyelitis. Marked disposition to pes varus.

My own examination elicited the following points: The boy was of stouter and shorter stature than his brother.

Intelligence good. His broad chest and fat stomach are in curious contrast to his spindle-shaped extremities. Circumference of chest, 26 inches. Right arm, 6½ inches; left arm, 6¼ inches. Right forearm, 6½ inches; left forearm, 6¼ inches. Grasp of both hands very weak. A general emaciation of all parts of upper extremities. Very distinct atrophy of infraspinatus. In the legs general atrophy is very well marked. The right thigh, four inches above patella, 11 inches; left thigh, 10¼ inches. Right calf, at greatest circumference, 8 inches; left calf, 8⅜ inches. The boy walks with a slightly waddling gait and has great difficulty in climbing the stairs. He can raise toes slightly on the left side, less well on the right side. Can raise left leg on tiptoe, but cannot do this with the right leg. In attempting to raise the whole body on tiptoe, falls forward. Sensation: Tactile sensation normal as determined by cotton, pin test, and the writing of numbers on the skin. Temperature sense normal. Pain sense exaggerated. Muscular sense normal. Plantar reflexes present and knee-jerk about normal. Slight lividity of legs, not so marked, however, as in the case of his brother.

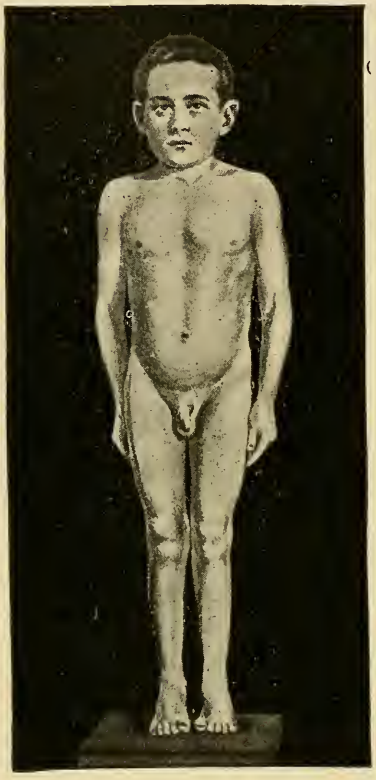


FIG. 85.
(Same patient as in Fig. 86.)

The electrical examination: In the upper extremities the faradic response in the median and ulnar nerves was decidedly diminished. In the median nerve first KCC with 13 MA; ACC not at 20 MA. Galvanic current: In the right leg no reactions could be obtained by excitation of the nerves with currents used. In the extensor hallucis longus the first KCC and ACC were

obtained with a current of 14 MA. The tibialis anticus did not respond to currents of 20 MA. The anterior thigh muscles and posterior thigh muscles respond to strong currents of 16 MA, without reversal of formula. In the left leg, the extensor hallucis, first KCC with 16 MA; first ACC with 18 MA. No contractions could be obtained by direct excitation of the tibialis anticus, with currents up to 20 MA; on excitation of the extensor digitorum communis there is a slight movement of the small toe.

The electrical examination, therefore, shows that the reaction of degeneration is present in its typical form in most of the muscles below the patella, the galvanic excitability of the peroneal nerve being entirely lost. It also shows changes in electrical behavior in nerves of the upper extremities, since the responses of the ulnar and median nerves were markedly diminished.

FIG. 86. — The Younger of the Two Brothers (see Fig. 84) after Second Operation, showing Correction of Deformity of Feet, Marked Atrophy of Legs, and Incipient Atrophy of Muscles above Elbow and around the Shoulder Girdle.

Comparing the histories of the two brothers, it was noted that they resemble each other very closely as regards the first appearance of the symptoms and the manner in which the disease spread from muscle to muscle; but there were also certain differences, such as the more marked electrical changes in the younger brother and the greater involvement of the upper extremities in him, than in the older boy. In the younger brother the disease was more fully developed in every respect than in his older brother; but such variations as occurred were within a reasonable limit and will serve to show to what extent variations may occur in persons undoubtedly suffering from the same type of disease.

In addition to the symptoms which these boys have exhibited, it is interesting to note that Vizioli has reported the occurrence of amaurosis in a similar case, due to an optic-nerve atrophy. This symptom would support the argument in favor of the nerve origin of the disease. Furthermore we may insist on the fact that hypertrophy has not been recorded in any case, and that fibrillary movements seem to occur in some.

Thomson and Bruce have reported an interesting case of a progressive muscular atrophy in a child; but they have not attempted to classify it under any special type. The disease began in the lower extremities, and gradually extended to the upper, involving both upper extremities and the muscles of the back. The gradual changes are well represented in Figs. 87–89. The child exhibited some disturbances of sensation (hyperalgesia); marked paresis in neck, back, and abdomen—most marked in loins, buttocks, and legs; least marked in shoulders and arms; no hypertrophy of any muscles. Electrical reactions at first little altered; later on faradic excitability considerably impaired; more markedly in the legs than in the arms. The report of the galvanic responses is too uncertain to permit of positive inferences; the disease was steadily progressive. The case is all the more unique, as the authors found a spinal lesion and only very slight changes in the peripheral nerves.*

ETIOLOGY.—Nothing more need be said upon this head than may be inferred from the previous histories. The disease is evidently a family affection, sometimes beginning at a very early age, as in my own cases, or appearing as late as the age of twenty, as in the one case described by Charcot-Marie. Whether the cases described by Osler as occurring in the Farr family of Vermont, which set in as late as the age of forty-six in some of the subjects, belong to this category or not is questionable, as the cases were reported before this special type of progressive muscular atrophy was known. In both my cases a thorough drenching of the skin by exposure to wet was mentioned, but I cannot attribute any further importance to this fact than that it may have helped to hasten a disease which was latent in the systems of the boys.

DIAGNOSIS.—The diagnosis of this "leg type," or neural form, of progressive muscular atrophy rests upon the recognition of the early beginning of the disease and of its hereditary or family character. Moreover, the paralysis beginning in the leg muscles, and spreading to the upper extrem-

*The case appears to bear a close resemblance to the peroneal form, as well as to an hereditary spinal form.

November, 1889.

January, 1890.

May, 1892.

FIGS. 87–89. — Case of Progressive Muscular Atrophy in a Child with a Spinal Lesion. (Thomson and Bruce.) The three figures illustrate the progressive wasting of the muscles and the deformities resulting therefrom. In the first cut, hyperextension of the fingers and of the big toe is very striking.

ities is associated with slight changes in sensation in the legs as well as in the arms. The occurrence of double club-foot (not congenital) will help to make the diagnosis still more certain.

The disease will have to be differentiated from hereditary ataxia, from chronic multiple neuritis, from poliomyelitis, and from the primary muscular dystrophies.

From hereditary ataxia the disease can be readily distinguished by the absence of the peculiar unsteadiness in walking and standing, by normal electrical reactions in cases of hereditary ataxia, and by the persistence of the reflexes in many of the cases of the peroneal form.

From chronic multiple neuritis we can distinguish the "leg type" by the fact that pain plays an even greater *rôle* in most cases of neuritis; that the atrophy is not steadily progressive, and that neuritis, of however long standing, rarely leads to double club-foot; and furthermore, that neuritis is not apt to occur as a family affection.

From poliomyelitis we can differentiate these cases by the very gradual development of the disease, in contrast to the more sudden onset in infantile spinal paralysis; by the progressive development of the atrophy, in contradistinction to the retrogressive character of the wasting in poliomyelitis. Poliomyelitis is not an hereditary or family disease, and if the wasting in a case of poliomyelitis is as great as in cases of the leg type of progressive muscular atrophy the knee-jerks and other deep reflexes will surely be lost, while in the leg type they may be preserved for a considerable period of time. It is not so easy always to distinguish these cases from the subacute forms of poliomyelitis; but if there is any doubt about the diagnosis in the earlier stages of the disease, the further progress of the trouble will remove all uncertainty.

The disease might also be confounded, in its later stages, with the Aran-Duchenne type of progressive muscular atrophy, particularly if, as sometimes happens, the atrophy in those cases attacks the legs very soon after it has begun in the upper extremities. Under such conditions we must rely for the differential diagnosis upon the fact that the Aran-Duchenne type begins, as a rule, much later in life; that it is rarely of an hereditary character, and that sensation is never affected in those cases as it is in the cases now under consideration. But I must concede that the clinical resemblance may be so strong between these two forms of disease that it will be practically impossible to differentiate between them.

PATHOLOGY.—For a number of years after this disease was first described the morbid anatomy was based upon mere speculation. Reference was made by several authors, among them by Hoffmann, to the older records of post-mortem examination by Virchow, Friedreich, and others. In these cases the authors found a degeneration of the nerves

and a degeneration of the columns of Goll, but no satisfactory statement could be made at that time, which antedated the discovery of modern staining methods, regarding the disease of the anterior ganglion cells. I objected to the use of these older records as a proof of the non-involvement of the spinal cord, but all doubt regarding the origin of at least some of these cases has been removed by the studies of Marinesco, of Dubreuilh (*Revue de Médecine*, 1890, p. 441) in a very typical case of a child dying, in a family of which the mother and eleven children were affected with the same trouble. Dubreuilh proves that there were old changes in the peripheral nerves, particularly in the motor nerves of the hands and feet, and that these changes diminished toward the spinal cord. The gray substance of the spinal cord was normal; there was a slight increase of the glia in the column of Goll, but not a true sclerosis, and the nerve-fibres were not diminished. The changes in the muscles consisted of simple atrophy of the fibres, of a loss of transverse striation, and of a proliferation of the nuclei. There were also some degenerated fibres and some in a condition of hypertrophy. It will be seen from this post-mortem account that the changes in the spinal cord were evidently secondary to those in the peripheral nerves; furthermore, that the changes in the muscles resembled more closely those occurring in the primary dystrophies than in diseases due to spinal processes. This form evidently holds a median position between the true spinal amyotrophies and the primary myopathies. (Cohn, Cassirer, and Oppenheim hold this same view.) The purely neural origin of the disease is in doubt. Later investigations may lend some coloring to the proposition of Bernhardt to speak of the cases as spinal neuritic atrophies, since the ganglion cells of the anterior horns, the anterior nerve-roots, and the peripheral nerves, after all, constitute a physiological unit. Further post-mortem examinations of the subjects of this disease may exhibit more considerable changes in the spinal cord than those which were found in Dubreuilh's case. I cannot consider the question settled, in spite of the autopsies recorded by Siemerling, Sainton, and others, and would urge a careful and detailed examination of every such case if opportunity presents itself.

The general treatment of these cases will be referred to in connection with the discussion of the other forms of muscular disturbance. I only wish to insist once more on the fact that the orthopedic surgeon may in these cases procure moderate relief, at least for a considerable period of time, by such measures as were employed by Dr. Gibney in the cases of the two boys under my immediate care.

PRIMARY MYOPATHIES (PRIMARY MUSCULAR DYSTROPHIES).

Our knowledge of these dystrophies is a recent acquisition for which we are specially indebted to the brilliant clinical and pathological studies of Erb. The German neurologist not only described a new form of progressive myopathy, but demonstrated very clearly the relations of this form to the older and well-known form of muscular pseudo-hypertrophy. In addition to this, Erb has subjected the innumerable types of muscular atrophy to a healthful criticism, and has thus helped to correct many erroneous views which were advanced by others. Excellent work has also been done by Charcot, Landouzy, Dejerine, Schultze, Strümpell, Dercum, Spiller, and others.

For a long time the Aran-Duchenne type of progressive muscular atrophy and muscular pseudo-hypertrophy were the only well-known forms of progressive muscular disease. With the evidence which proved that pseudo-hypertrophy was never of spinal origin, a wide distinction was created between the spinal forms and the primary myopathies. As new types of muscular diseases were described by individual authors the question arose in each instance, whether the cases reported inclined rather to the spinal than to the pseudo-hypertrophic form. It is only very recently that sufficient evidence has been brought forth to show that all the primary myopathies are closely related to one another, and that the types that have been so carefully described in former years, are practically nothing but peculiarities in the topographical distribution of diseases which should be included under the broad term of progressive muscular dystrophies. Much was at one time made of the occurrence of atrophy or hypertrophy, but less importance is attached to this point now, for we know that the hypertrophic stage represents in most instances an earlier stage of the disease, and that atrophy rapidly supervenes upon this apparent or real hypertrophy of muscular tissue. Hypertrophy never occurs in the spinal forms of muscular wasting,* and in this respect it

* Single hypertrophied fibres have been found in sections of muscles from cases of progressive amyotrophy.

is a most significant symptom, but its occurrence is not a sufficient basis for a classification of the various types of primary myopathy.

Before entering upon a detailed account of the primary myopathies, it will be well to state the chief features of the various types. By placing the symptoms in parallel columns the entire subject can be understood more readily:

TYPES OF PRIMARY DYSTROPHIES.

	MUSCULAR PSEUDO-HYPERTROPHY.	JUVENILE FORM OF PROGRESSIVE MUSCULAR ATROPHY (ERB'S TYPE).	TYPE LANDOUZY-DEJERINE. ("INFANTILE FORM.")
Part first affected.	Legs (calves).	Shoulder girdle.	Face and shoulder-girdle.
Distribution of hypertrophy.	Calves, rarely thighs.	Muscles around shoulder-girdle and pelvic girdle.	None.
Distribution of atrophy.	Thighs, deep muscles of back, shoulder, and scapular muscles. Calves during later period; at that time also general atrophy.	Thighs, deep muscles of back, upper arm. Hypertrophied parts may become atrophic in later stage.	Face muscles, including lips and orbicularis palpebrarum; shoulder and scapular muscles.
Parts remaining normal.	Face, forearm, and hand, except in last stages.	Face, forearm, hand and leg muscles, except in last stages.	Forearm, hand and legs, and deep muscles of back.

If we add to the above table the facts that in all these three forms there is a distinct history of heredity, or at least of the occurrence in various members of the same family; that there are often slight and sometimes very marked quantitative changes in the electrical reactions, but that there is rarely if ever a complete reaction of degeneration; if we note, furthermore, that the reflexes may remain preserved for a considerable period of time and then disappear, in keeping with the progress of the muscular wasting, we shall see that there is practically no other distinction between these various types of myopathies than the mere distribution of atrophy or hypertrophy. The hereditary type of progressive muscular atrophy as it was described by Leyden, is not included, simply because heredity is not a sufficient basis of classification, and many of his cases would belong more properly either under the heading of Erb's type, or under that of the peroneal form of progres-

sive muscular atrophy. There appears at first sight to be a broad gap between the cases of muscular pseudo-hypertrophy and the cases of Erb's type, but these different types may be represented in various members of a single family. The relation of these types to one another is convincingly demonstrated by the cases of three brothers to be referred to in this chapter. (Figs. 90–92.)

FIG. 90. — Oldest Brother, aged Sixteen Years, exhibiting Atrophy following Pseudo-Hypertrophy of the Calves, and Extreme Atrophy of Shoulder Muscles. Boy unable to move from chair or to hold himself erect (late stage of "pseudo-hypertrophy"). Figs. 106–108 represent three brothers.

Landouzy-Dejerine have insisted on the right of their type to special consideration, claiming that ordinary muscular pseudo-hypertrophy, and even Erb's type of disease, were never associated with an involvement of the face, yet it is very certain that their type is practically nothing more than that described by Erb, plus involvement of the face muscles. Landouzy-Dejerine also denied that an atrophy of the face muscles was ever associated with typical pseudo-hypertrophy; but Westphal first published a case which showed that the face muscles are occasionally affected in cases of typical pseudo-hypertrophy, and I was able, some years ago, to record a case which was a typical representative of muscular pseudo-hypertrophy, which passed through the stage defined by Erb's type, and in which the muscles of the face were also involved. There is, therefore, no sufficient reason to retain the Landouzy-Dejerine type as a separate form of disease.

The characteristic symptoms of the various types may now be described.

MUSCULAR PSEUDO-HYPERTROPHY.—As described many years ago, above all by Meryon, Duchenne, and Gowers, pseudo-muscular hypertrophy is characterized by its occurrence in early youth. Boys are affected somewhat more frequently than girls; but although affecting boys, the disease is inherited almost invariably through the mother. The first symptoms are, a weakness in the muscles of the leg and an early increase in the size of the calf muscles. In rare instances the hypertrophy may begin in the thigh muscles. The gait is waddling, and the child soon finds difficulty in walking up and down stairs, in climbing on chairs, in rising from the floor or from any recumbent posture. In the earlier stages of the disease the patient rises from the floor by dint of great effort (see Fig. 109) and by "climbing up upon himself." In later stages of the disease the patient, if put on the floor, lies absolutely prostrate and is not even able to raise the head from the floor. Sitting up without support may be entirely impossible. As the weakness and atrophy increase, the patient becomes more

FIG. 91. — Second Brother, aged Thirteen and a Half Years, exhibiting Hypertrophy of Calves, of Gluteal Muscles, and of Muscles about the Shoulder Girdle; Distinct Atrophy of Arm Muscles (Erb's Type, or Juvenile Form of Progressive Muscular Atrophy). All the muscles are now beginning to atrophy; boy is only a little less helpless than his older brother.

and more helpless, is unable to stand or to walk, becomes either bedridden, or is compelled to sit in a chair and even loses the use of the upper extremities; is not able to raise

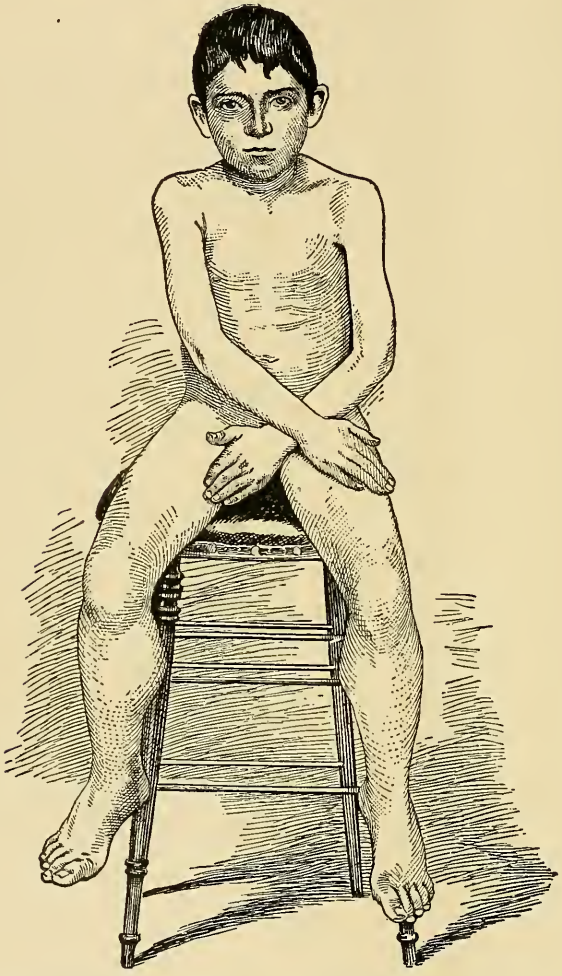


FIG. 92.—Youngest of the Three Brothers, in the Earlier Stage of Pseudo-Hypertrophy. (For the photographs of this boy I am indebted to the courtesy of Dr. Collins.)

the arms, and may have no use of any of the muscles, except the small muscles of the hand. In the two boys (Figs. 90, 91) under my observation, a *climbing up of the hand along*

the head in order to get the arm into the erect position was a very characteristic feature.

In addition to the hypertrophy of the calf muscles, we now know that there is apt to be atrophy of the muscles of the thigh, of the arm, and the shoulders; the scapular mus-

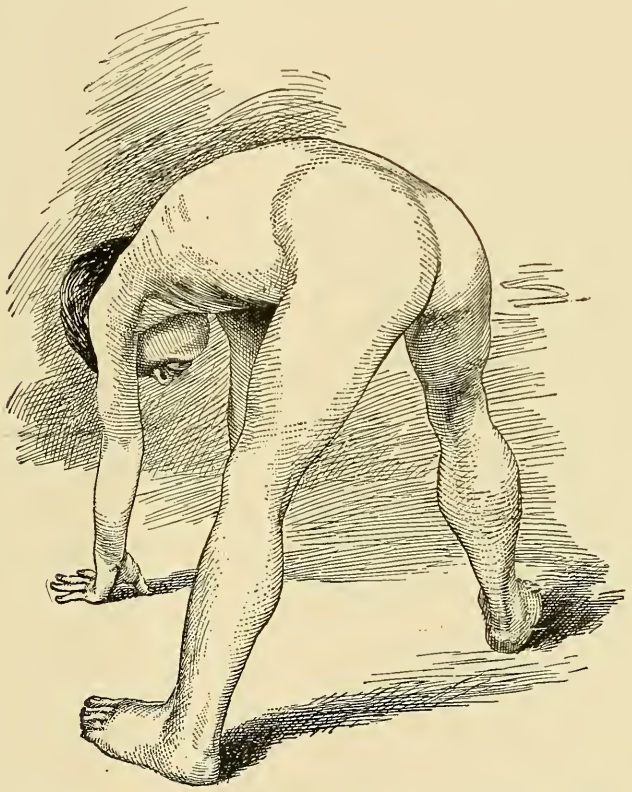


FIG. 93.—Boy with Pseudo-Hypertrophy attempting to straighten himself. Same patient as in Fig. 92.

cles, and at a very early stage of the disease the serrati, the latissimus dorsi, and the pectoralis major, are often wasted. The forearm muscles and the hand muscles are rarely affected. The disease is often associated, as the other forms of myopathy may be, with symptoms of a general degeneration; thus I have found nystagmus, lisping speech, and a moderate degree of imbecility in not a few of these cases.

Erb's Type, or the Juvenile Form of Progressive Muscular Atrophy.—According to Erb's own summary, this type is characterized by progressive wasting with weakness of certain groups of muscles, beginning either in childhood or early youth, involving, as a rule, the muscles of the shoulder girdle, the upper arm, the pelvic girdle, the thigh, and the back; the forearm and leg muscles remaining intact for a very long time. The atrophy may be associated with true or pseudo-hypertrophy of some muscles. The pectorals, trapezii, latissimi dorsi, the serrati, the rhomboids, as well as most of the upper-arm muscles and supinators are apt to be wasted; while the deltoids, supraspinati, and infraspinati are either normal for a long time or hypertrophied. There are no fibrillary contractions and no reaction of degeneration; no sensory or visceral disturbances. (See Fig. 91.)

Fig. 94.—Patient with Landouzy-Dejerine Type. Indication of bouche de tapir; patient cannot show upper teeth, nor close eyes. (See also Fig. 8.)

A few years ago, I was inclined to consider Erb's type a great rarity in this country; it is unquestionably the least frequent of all the forms of progressive muscular atrophy which we have occasion to see in clinics or private practice; but I have seen at least a dozen cases of this form within a period of five years, from which the reader may gather the frequency or infrequency of the disease; but there is no doubt that many cases exist which have not been reported, simply because they have not been properly recognized.

The Facio-Scapulo-Humeral, or Landouzy-Dejerine Type.—This type includes cases in which the atrophy begins early in life, and, as a rule, in the muscles of the face, giving rise to what the authors have termed the "*facies my-*

opathique;" the lips are considerably thickened and constitute the *bouche de tapir*, or tapir mouth. Later on in the course of the disease the atrophy spreads to the shoulder and arm muscles. The supraspinati and infraspinati, the subscapularis, and flexors of the hands and fingers remain normal. Among these muscles that remain normal it may at once be noted are several which are distinctly hypertrophied in Erb's type. In the Landouzy-Dejerine type the muscles of deglutition, of mastication, the respiratory and laryngeal muscles, as well as the ocular muscles, remain normal. In exceptional cases the disease may begin in the shoulder or arm muscles, or even in the lower extremities. The disease is distinctly hereditary. Fibrillary contractions and reaction of degeneration are never present.

I have given these symptoms as nearly as possible as they were stated by the authors themselves in order to do full justice to their cause, but it will be evident, without further argument, that, with the exception of the involvement of the face, there is very little distinction between these cases and those of the juvenile form of progressive muscular atrophy.

The various types of progressive myopathy are sufficiently illustrated by Figs. 90–94, which supply additional evidence in favor of the intimate kinship existing between these various types.

Figs. 90 and 91 represent two brothers who had been under my observation for many years; the one is an example of muscular pseudo-hypertrophy with atrophy of shoulder and trunk muscles; the other boy's disease began as a muscular pseudo-hypertrophy, but the hypertrophy of the muscles above the shoulder and pelvic girdles constitute it a most pronounced case of Erb's type—the juvenile form of progressive muscular atrophy. A third brother was seen in the state of pseudo-hypertrophy (Fig. 92). After a lapse of years the pseudo-hypertrophy may disappear, and all three brothers will then bear the closest resemblance to each other. Fig. 94 is a photograph of the patient afflicted with the Landouzy-Dejerine type of muscular atrophy.

To these I wish to add a brief history (published in 1890), of a case which was one of unusual importance, as the patient presented the combined symptoms of all known types of primary myopathy. A young man, twenty years of age, whose history is entirely negative, has noted, since early childhood, a peculiarity about his face; he was not able to whistle as well as other boys,

and as long as he can remember his face appeared twisted to one side, at least while speaking. It was not until one year before my examination that he became aware of any further trouble. He claims to have struck his shoulder, and since that time to have noticed a weakness of both upper extremities. He was employed by a surgical-instrument maker and had to lift heavy boxes, but this he is no longer able to do.

During the examination, was asked to whistle; he could not do it, nor could he keep his eyelids tightly pinched. The condition of his muscles may be summarized as follows:

Wasted: Both pectorals, major and minor. Both serrati, right more than left. Both latissimi dorsi, left more than right. Levatores anguli scapulæ, right more than left. Rhomboids, left more than right. Both trapezii (middle and lower third), left more than right. Both biceps muscles and both brachiales-antici. Both triceps muscles, right more than left; the supinator longus of both arms, right more than left. Anterior thigh and leg muscles, left more than right. Posterior thigh muscles (thin). Orbicularis palpebrarum of each side and the orbicularis oris.

Normal: Back muscles, forearm and hand muscles, gluteal muscles, and muscles of foot.

Hypertrophied: Deltoids, infraspinati, supraspinati, and calf muscles, but the right calf is beginning to waste.

There were no fibrillary contractions in any of the affected muscles. All the muscles, including those wasted and those hypertrophied, responded to both currents in proportion to the quantity of normal contractile fibre that each muscle retained. There was no reaction of degeneration in any muscle. The knee-jerks were present. There was no ataxia, no disturbance of sensation, and there was not a single symptom pointing to an involvement of the central nervous system.

From the condition of the calves, there might be some reason to class this case among the pseudo-hypertrophies; from the appearance of the shoulder-girdle and the thinness of the upper arm, we might rank it with Erb's juvenile form; and if we take the face into consideration, we might classify it with the Landouzy-Dejerine type of progressive muscular atrophy. It does not quite tally with the older accounts of pseudo-hypertrophy; for although the shoulder muscles are sometimes involved in such cases, it is exceptional to have both shoulder and face muscles affected. From Erb's form it is distinguished by the involvement of the face muscles, and from the ordinary cases of the Landouzy-Dejerine type this case is to be distinguished by the additional involvement of the calves. It will not do to suppose that the boy is affected with three different diseases; it is much more to the point to state that the symptoms in this case prove that the three distinct forms practically represent subdivisions of one and the same disease, and that the primary muscular disease was so fully developed in this patient that he practically represented all the known types of progressive myopathy. We see by this case, too, how wrong it is to make too much ado over the varying distribution of atrophies or hypertrophies.

DIAGNOSIS.—It is much more difficult to differentiate between the various forms of progressive muscular dystrophies than to distinguish between them and other diseases. The factor of heredity, the occurrence in families, the absence of fibrillary twitchings and of changes in electrical reaction, and above all the slowly progressive character of these diseases and their onset in early life, will scarcely permit of any confusion with other diseases. If the patients are examined later in life, after extensive atrophy has supervened upon preceding hypertrophy, if the atrophic paralysis is so extreme that the patient is bedridden and that there are practically no muscles which respond to the electrical current, if all the reflexes are absent, there may be considerable difficulty in differentiating between such a condition and that of chronic poliomyelitis. But even at such an advanced period of the disease the former history of the patient will help to establish a correct diagnosis.

Mention should, however, be made of one other condition which I have met with but twice, in which the question has come up whether the patient was suffering from a form of primary muscular dystrophy. Both of these cases were instances of what I would wish to call physiological hypertrophy. The one was the case of a physician who had attained unusual muscular development in his efforts to correct phthisical tendencies, and the second was the case of a brother of a well-known physician who, through inordinate exercising at lawn tennis had produced a hypertrophy of the shoulder, arm, and forearm muscles of the right side. In both these cases I was consulted because of a weakness which had followed upon this unusual hypertrophy. In the case of the physician first referred to, several muscles (among them the infra and supra spinati, and the deltoid) had begun to atrophy distinctly. The possibility of the development of some form of progressive muscular atrophy was entertained by others in both cases, but I was certain that this grave prognosis was not justified, for the entire development of the trouble, the onset late in life, and the occurrence of these conditions in persons with a clear family record furnished the best evidence that the condition of these two patients was the result of over-exercise and nothing more.

The point of greatest interest in both these cases is the surprising weakness of the muscles in spite of the hypertrophied condition. It would seem that any muscular fibre forced to an unnatural growth (hypertrophy) is likely to succumb in the struggle. In both these cases I insisted on absolute rest of the affected parts and on the use of common sense in physical exercise. In the case of the physician all the muscles have returned to a normal

state and have remained so for fully three years; in the second case the normal strength of the arm has returned, and there is no sign of an impending atrophy.

The points of differential diagnosis between the spinal forms of progressive muscular atrophy and the primary dystrophies have been insisted upon over and over again. The rule is that in spinal cases the affection is not hereditary and generally begins in the upper extremity; there are fibrillary contractions, and marked changes in electrical reaction; while in the dystrophies heredity is the most prominent factor, the diseases begin early in life, there are no fibrillary contractions, and the electrical reactions remain normal or nearly so throughout the entire course of the disease until the stage of extreme atrophy is reached.

After the consideration of these cardinal symptoms there should be no real difficulty in distinguishing between these two principal forms of progressive muscular wasting; yet cases appear every now and then in which the symptoms are so distributed that it is impossible to classify them by adhering to these cardinal symptoms. The most interesting case in point is unquestionably the one reported by Strümpell. In this patient the disease did not begin until the age of twenty-nine, in the fingers of the right hand, but there was a strong history of heredity, his mother having suffered from a similar disease. The atrophy spread from the small muscles of the right hand to the muscles of the shoulder, and later on to the muscles of the opposite arm. The deep muscles of the spine, the glutei, and the thigh muscles remained entirely intact, after a period of more than eleven years. The symptoms just stated pointed to a spinal form, rather than to a pure myopathy; the heredity was, however, more in keeping with the primary dystrophies, and the entire absence of fibrillary movements as well as of changes in the electrical reactions would have inclined one to place the case in the category of primary myopathies. The histological character of the muscular tissue was also more like that described in cases of primary myopathies, yet at the autopsy decided changes were found in the spinal cord, in the anterior nerve-roots, and in a number of the peripheral nerves. Thus the impropriety of classing such a case exclusively under one heading or the other was clearly demonstrated, and I think Strümpell quite right in insisting that the chief value of his case was in showing that these cases of progressive muscular atrophy, whatever form they take, belong to the order of hereditary systemic diseases, and that it is largely a matter of chance or else due to causes still unknown whether the peripheral or the more central portion of the second division of the motor tract becomes the chief seat of the disease. A case of Savill's shows that the symptoms of an amyotrophy may coexist with those of a myopathy of the Landouzy-Dejerine type.

PATHOLOGY.—The designations, "primary myopathy," or "primary muscular dystrophies," which have now been universally accepted for the diseases under consideration, are meant to imply that the origin of the disease is in the muscular system itself, and that it is not due to changes either in the peripheral nerves or in the spinal cord. With regard to the more recently described forms of primary muscular dystrophy, there has never been any doubt as to the non-spinal origin, but a long and hot discussion was waged over the spinal origin of muscular pseudo-hypertrophy. In 1888 I analyzed all the cases of muscular pseudo-hypertrophy that were accessible at the time, and found that of twenty-five cases which had been reported, eight had to be excluded because the spinal cord was not examined microscopically, or because the examination was not properly made. Of the seventeen remaining cases, the spinal cord and anterior nerve-roots were found absolutely normal in twelve, and in five others the changes that were found could not be held responsible for the changes in the muscles. A similar conclusion has been reached by other authors, and at the present time no author of repute has ventured to fall back upon the older theory of the spinal origin of muscular pseudo-hypertrophy. Such slight changes as were observed by some—the diminution, for instance, in the number of ganglion cells or slight changes in the contours of these cells—can now be sufficiently explained on the supposition that all such changes are secondary to the peripheral muscular trouble.

The histological condition of the atrophied or hypertrophied muscles has been studied with particular care, for it was supposed for a very long time that the diagnosis of a myopathy or of an amyotrophy could be based safely enough upon histological appearances; but we shall see that this hope of finding some absolute point of differentiation between these two conditions can no longer be entertained. In muscles atrophied from spinal lesions, the following were supposed to be the chief changes: a loss of striation of the muscular fibres and a narrowing of the same, an increase in the number of muscle nuclei, and possibly segmentation of the nuclei, granular or fatty degeneration of the fibres,

and occasionally globules of fat between the muscle fibrils. Some or all of these changes are present according to the length of time that the atrophy has existed, but all of these changes have also been found in spinal forms, as well as in primary dystrophies. The increase in the nuclei is not so great in the dystrophies as in the amyotrophies. In the spinal forms it was thought that hypertrophied fibres were never found, while they are extremely common in the purely muscular types. But Mueller and others have recorded exceptions to this rule and have proved the presence of hypertrophied fibres even in cases of poliomyelitis of old standing. A large number of hypertrophied fibres in a given section of a muscle is, nevertheless, more frequent in the primary muscular dystrophies than in the spinal atrophies.

The next question that arose was whether it was possible by histological examination to differentiate between the various primary myopathies. In muscular pseudo-hypertrophy we have, as a rule, a narrowing of the fibres with changes in their contour, granular or fatty degeneration of the fibrils, and accumulation of fat globules between them, and increase, without marked proliferation, of the connective tissue. Hypertrophied fibres can be found scattered in between fibres of normal dimensions, or between those that have evidently undergone atrophy. A slight increase in the muscle nuclei is often found, but unusual increase is suspicious of a spinal origin. Jacoby thought that the disease consisted in the main of a chronic inflammation invading both the perimysium and the muscle tissue, and was inclined to term the process a myositis progressiva hyperplastica, but his views and his histological findings have not been corroborated by others. Westphal, in a very typical case of pseudo-hypertrophy, found, on post-mortem examination, enormous increase of adipose tissue in which the muscle fibrils were nearly of normal size, increase of the interstitial connective tissue, no hypertrophied fibres, and strands of connective tissue occasionally passing through the fatty parts; a few of the groups of muscle fibres appearing to be strangulated by the strands of connective tissue. In sections from a case which I have recently had occasion to examine, the fibres were found to be of varying size and there was a distinct proliferation of the muscle nuclei. Schultze, in a case which stands midway between pseudo-hypertrophy and Erb's juvenile form, found, in addition to peculiar giant-cell formations, a large number of fat-cells in the muscular tissue, an increase of the connective tissue, and remnants of hypertrophied, normal, and atrophic fibres, and an enormous increase of nuclei, which the author thought greater than in the ordinary cases of pseudo-hypertrophy. He also described the occurrence of vacuoles which were in all probability not due to the hardening

process. Hitzig's observations were of special interest for a time, as he examined four cases most carefully. He concluded that "the primary and most important change in juvenile atrophy is not an interstitial process, but decidedly parenchymatous, and according to the intensity of the disease is represented by slight or excessive hypertrophy of the fibres . . . The anatomical changes in pseudo-hypertrophy, on the other hand, are characterized by active changes in the connective tissue." This distinction between the histological changes in pseudo-hypertrophy and Erb's form of primary myopathy has not been borne out by others, and if I am not mistaken, Hitzig has withdrawn his former views. The matter was definitely settled by more recent studies of Erb, Cramer, Pick, and others (Figs. 95 and 96). Erb proved that in all cases of progressive muscular dystrophies, whether of one type or another, the changes in the muscular tissue were very similar, and that such changes as did occur were simply due to different stages of the disease. Erb showed (Fig. 95) that hypertrophy and atrophy of fibres, marked proliferation of the nuclei, vacuolization (near *b*), and segmentation (*c*) of fibres were the chief changes, and that these occurred in all possible forms of primary dystrophies, and of these phenomena the hypertrophy of the fibres seemed to be the first to appear.

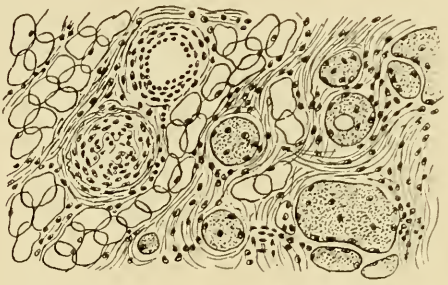


FIG. 95.—Changes in Muscular Tissue in a Case of Primary Dystrophy. (Erb.) *a*, Above altered blood-vessel; for other lettering see text.

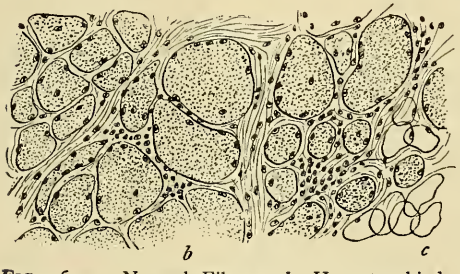


FIG. 96.—*a*, Normal Fibres; *b*, Hypertrophied Fibres; *c*, Atrophied Fibres and Fat. (Erb.)

From a case of the Landouzy-Dejerine type (Fig. 97), I excised a large piece of the infra-spinatus muscle, and in doing this a piece of the nerve as it enters the muscle was accidently removed. On histological examination of the specimen, prepared for me by Dr. Wiener, we found a considerable amount of fatty tissue between and around the muscular fibres (Figs. 97, 98) much atrophy, but no hypertrophy of the muscle fibres; there was also distinct evidence of a marked degeneration of the nerves. The finding was a very singular one, but it will not do to draw too broad an inference from a single case. At all events, the participation of the nerve (whether in primary or secondary fashion) in a disease supposed to be a purely muscular affection is worthy of note.

After a very long discussion on the histological changes in progressive muscular dystrophies, Erb concludes that

FIG. 97.—Section of Infra-Spinatus Muscle from a Case of Landouzy-Dejerine Type, showing Deposits of Fat and Degenerated Bundles of Nerves. (Low power. Drawing made after specimen stained according to Van Gieson.)

these dystrophies are forms of a tropho-neurosis, which may occasionally be the result of functional disturbance of trophic centres; and that such disturbance may be either

FIG. 98.—A Part of the Nerve Bundles more highly magnified, showing Degeneration of Nerve Fibres. (See preceding figure.)

primary or secondary to spinal lesions—a conclusion not unlike that reached by Strümpell a few years later in the article referred to above.

DURATION.—All these forms of muscular dystrophies take a relatively slow course, but the period of time that elapses before the stage of utter helplessness is reached varies very much in different individuals. It is safe to say that those suffering from muscular pseudo-hypertrophy are the ones most apt to be completely crippled at an early day. Two or three years often suffice to make the patient entirely helpless. Erb's juvenile form and the Landouzy-Dejerine type progress more slowly —a number of years elapsing before the legs become involved. I have seen one case of Erb's form which has lasted over thirty years, and another of the typical muscular pseudo-hypertrophy, which has lasted at least fifteen; but if death does not ensue from the disease itself, patients may be carried off by any slight intercurrent disease.

FIG. 99.—Boy with Defective Development of Scapula and Shoulder Muscles.

TREATMENT.—Interesting as these diseases are from a medical and scientific point of view, they are unfortunately most unsatisfactory as regards the results of treatment; and yet there is no need of despairing and declaring that nothing can be done in the earlier stages of these troubles. Contractures of muscles and deformities of various parts are extremely annoying and prevent the patient from getting about freely, or without the use of crutches or wheeling-chairs. I wish to plead for the early and active interference of the orthopedic surgeon. As the diseases are at times but very slowly progressive, the correction of deformities by surgical measures may bring relief for a considerable period of time. Excellent though temporary

results were obtained in several patients whom I have treated in conjunction with Dr. Gibney. His results in the cases of the peroneal form of progressive muscular atrophy make one a little hopeful that still better results may be attained in the future in this and other forms of muscular atrophy by similar or even bolder measures; but as Oppenheim has recently insisted, such cases are not suitable for tendon transplantation. Intelligent exercise of atrophied or hypertrophied muscles, diligent and prudent use of massage and electricity in these cases will be in order. The latter agent has a wholesome influence upon the condition of muscles whose nutrition has been altered, and is furthermore a convenient form of moderate exercise. In the case of a boy suffering from the Landouzy-Dejerine type, all the symptoms were brought to a standstill, and a decided improvement has been effected by the careful administration of electricity. If the stage of atrophy and helplessness has been reached, nothing more can be done than to make the patient as comfortable as possible.

FIG. 100.

TOTAL ABSENCE AND EARLY ATROPHY OF MUSCLES.

This condition is suggested by the preceding discussion of the primary muscular dystrophies, and may as well be referred to in this connection as in any other. The condition is a rare one. The following case, which I have seen, suggests a similar condition, and at the same time shows

that the defect may include bony parts as well as the muscles covering them.

The boy whose condition is represented in Figs. 99 and 100 was seen by me a few years ago. He was then six years of age. The mother stated that she had noticed during the past year that the right shoulder was higher than the left. On examination we found that there was shortening of the right scapula, the longest diameter being 4 ctm. less than that of the left, and there was insufficient development of the right supra- and infra-spinati of the latissimus dorsi, and of the pectoralis major. It will be noted that these are the very muscles which are often involved in cases of primary muscular dystrophies. The condition is of some interest as showing that the muscles which are most subject to disease are also found congenitally defective. Whether this defect of development is due to some peripheral nerve anomaly, or whether it is the result of defective development of the spinal-cord cells, cannot be stated.

Dejerine and Thomas have described a form of disease which they designate as an "hypertrophic and progressive interstitial neuritis of childhood." It bears some resemblance to the peroneal form of progressive muscular atrophy, and to hereditary ataxia. The chief symptoms are: Progressive wasting of the muscles at the distal ends of the extremities, lightning pains, Romberg symptom, contracted pupils, sluggish reaction of pupils or complete immobility, nystagmus, diminution of electrical responses, hardening of nerve trunks as determined by palpation. Anatomical findings were: Chronic interstitial neuritis, most marked at the periphery; sclerosis of the columns of Goll and of Burdach; atrophy of the anterior horns and anterior roots.

CHAPTER XXII.

MALFORMATIONS AND CONDITIONS DUE TO DEFECTIVE DEVELOPMENT OF THE CORD.

ARREST of, or disturbance in, the development of the spinal cord is generally associated with similar conditions affecting the brain. These spinal-cord conditions have, with the exception of spina bifida, very little clinical interest. The chief malformations are the following:

AMYELIA, OR ENTIRE ABSENCE OF THE SPINAL CORD, is associated with a condition of anencephaly, and the absence of both may be designated as amelyencephaly. In some instances there is tolerably normal development of the cord, however, while every trace of the brain may be wanting. This defect of the spinal cord is in some instances a simple form of agenesis, in other cases a start has been made in the development of the central nervous system, but an unusual accumulation of cerebro-spinal fluid at a later period of fœtal life has destroyed the neural axis. In forms of amyelia there is at times also a defective development of the vertebræ. If the defect is due to a spinal dropsy we are able to trace, after the birth of the child, the vertebral canal and the spinal membranes which are distended by the excessive accumulation of fluid.

ATELOMYELIA is the term by which the older authors designated a partial lack of development, such as the entire defect of the pyramidal tracts, or a slight defect in the substance of the cord which occurs at the site of the spina-bifida. A similar defect is at times observed in the lower portions of the spinal cord, which is generally associated with some congenital defect in the development of the lower extremities. The only form of atelomyelia which is of any practical significance, is the one referring to the defective development of the pyramidal tracts, and that has been considered in the chapter on hereditary spastic paralysis.

ASYMMETRY OF THE CORD has been described in a few instances. It is probably due to defective development of some of the systems of the spinal cord, particularly of the pyramidal tracts on one side of the cord, and not on both.

HETEROTOPIA is a condition which has aroused considerable interest of late. It denotes malposition of the gray matter of the cord; parts of which are found scattered among the systems of white fibres, either in the lateral

columns or even in the posterior columns. Van Gieson has shown that heterotopia can easily be caused by careless manipulations of the spinal cord during post-mortem removal from the body. He throws some doubt upon the existence of any such condition as heterotopia during life. While it may be conceded that many of the cases that have been reported as heterotopia, were, in all probability, artificially produced, we cannot deny the occasional existence of such a condition: for it has been found when the spinal cord has been most carefully removed, and, furthermore, if it were invariably an artificial condition, we would surely find it in many more spinal cords than is actually the case.

A small cord or *micromegaly* has often been noted; in cases of hereditary cerebellar ataxy for instance. A normal adult cord has a diameter of 6 to 9 mm. in the dorsal vertebræ; 8 to 11 mm. in the upper cervical; 15 mm. in the cervical, and 12 mm. in the lumbar enlargement.

DIPLOMYELIA denotes a double cord. This condition is at times found in monstrosities of various kinds, but occasionally in children who have exhibited no spinal symptoms during life. It is due to a defective development of the spinal cord, and is to be attributed chiefly to an abnormal dilatation of the central canal. In the first months of fœtal life the central canal is relatively wide, and if normal development takes place the canal becomes smaller and smaller as time goes on. If this retraction does not take place, the substance of the cord remains divided and we practically have a double structure. This division is, however, never complete, except in a few cases associated with anencephaly. This diplomyelia need not involve the entire cord. At times it is restricted to a few segments only. A double central canal is not a great rarity. It denotes an arrest in the normal development of the cord and as a rule involves only a part of the medulla spinalis. The two canals lie side by side. The relation which these defects of the central canal bear to syringo-myelia is very evident and need not be dilated upon. But it will be remembered that there is some doubt whether syringo-myelia represents an arrest in the development of the spinal structures, or a destruction of parts that have been normally developed. If the ependymal lining of the central canal is entirely normal the syringo-myelia is likely to be a congenital condition and not one due to disease. The distention of the central cavity has been described as hydrorrachis interna. In contradistinction to this we have the condition of

EXTERNAL HYDRORRACHIS AND SPINA BIFIDA. — This is by far the most important of all the congenital anomalies of the spinal cord, and is by no means a great rarity, occurring in about one of every thousand cases. Spina bifida denotes a condition in which there is an unusual accumulation of serous fluid in the vertebral canal either between the pia mater and the arachnoid or between the arachnoid and the dura. Associated with this in-

crease of fluid is a cleft in the vertebræ due to an absence of the vertebral arches, and in some instances the bodies of the vertebræ are wanting. If there is no external prominence the condition is called spina bifida occulta. Recklinghausen insists that the dura also is cleft in the majority of cases. Through this cleft in the vertebræ the spinal membranes or the cord may protrude, and according to the parts protruded we can distinguish three kinds of spinal hernia: First, meningocele; second, meningo-myelocele; third, syringo-myelocele. The first two are much more common than the last named. The annexed figures will illustrate the condition better than any verbal descrip-

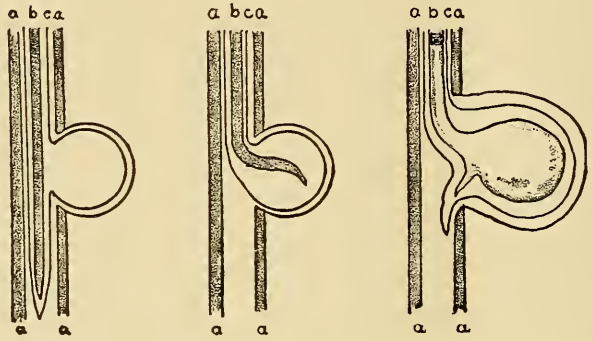


FIG. 101.—Meningocele, Meningo-myelocele, and Syringo-Myelocele. *a*, vertebral walls; *b*, cord; *c*, membranes. (Dana.)

tion would. In all of the cases the wall of the sac is lined by the arachnoid, but not invariably by the dura. The pia is not part of the sac-wall unless there is also an internal hydrorrhachis or hydromyelus. In some of them the spinal cord as well as the nerve-roots protrude into the sac. In others again only a few nerves are found in it, the tumor contains cerebro-spinal fluid, some connective tissue, and, as a rule, considerable fat.

ETIOLOGY.—Spina bifida is caused by defective closure of the vertebral arches, formed from the mesoblast.

This is a primary defect in development, and not caused by dropsy of the spinal cord, as was once supposed to be the case.

Normally the vertebral column is closed from above

downward. It is natural, therefore, that the defect should be found most frequently in that part (the lumbar segments) which is the last to close. The external tumor is generally to be seen in the region of the second to fifth lumbar vertebræ. There is distinct fluctuation in the majority of cases; the size may vary from that of an egg to the size of a child's head. The skin of the tumor may be normal; in some cases it is thinned out, particularly if the tumor grows rapidly. Ulcers are occasionally formed; the skin may be ruptured, but the dura continues to act as the wall of the sac. In many cases there is a narrow opening between the sac and the spinal canal. At times there is a complete constriction at this point, giving rise to an ordinary cystic formation which may contain some of the spinal structures that have been separated from the main contents of the spinal canal.

FIG. 102.—Child of Seven Years, with Spina Bifida and Deformity of the Feet.

SYMPTOMS. — The chief symptoms are those of increased pressure in the spinal and cranial cavities, and such as point to a direct interference with the functions of the spinal cord. Children with spina bifida are often poorly developed, and mentally deficient. If the child lives long enough it presents spastic or flaccid paralysis of the lower extremities with anæsthesia and with or without atrophy. Talipes varus is frequent. Remak described a form of club-foot associated with spina bifida, differing from congenital club-foot. The child is late in learning to walk and has little or no control over the sphincters of the bladder and rectum. Ectopia of the bladder and other viscera, defective development of the

lower extremities, are frequent complications of spina bifida.

DIAGNOSIS.—The condition is easily recognized; it is to be differentiated from congenital and other tumors which occur often enough in the lumbo-sacral region and may be connected either with the substance of the cord or with the spinal membranes.

Fibroma, sarcoma, myxoma, hygroma, and echinococcus cysts are the forms of tumor which occur commonly in this region. It is of importance to decide whether the sac contains nerve structures or fluid only. An exploratory puncture with a needle (attached to an electric battery) will help to clear up the point.

The course and prognosis of spina bifida will vary much according to the extent of the hernia; in some instances, the sac ruptures before or very soon after birth; these cases are invariably fatal. The danger of rupture with subsequent purulent meningitis is great in all cases. If the opening of the sac is closed, the child may continue to live for a long period, but it is after all rare for a person with spina bifida to live to middle life. If the sac contains nerve structures, the prognosis is less favorable than if it contains fluid only.

TREATMENT.—Much ingenuity has been exercised by surgeons in the attempt to cure these cases. Compression and ligature (of the pedunculated tumor) have been practised in former years, but the results have been disappointing. Ahlfeld was probably the first to cure a case of this sort by surgical procedure. Of late years surgeons have become much bolder. Bayer and Hildebrand, Schede, Bergmann, and others have reported a number of successful cases. The meningoceles and the meningo-myeloceles are considered fit cases for operation if the child's health is good, and if there are no serious complications such as hydrocephalus, deformities of the extremities, ectopia of the bladder and the like. In case the exact contents of the sac are in doubt, the X-rays may be used. A free incision of the sac is always to be preferred to injections into it of iodine and similar fluids.

DISEASES OF THE BRAIN.

CHAPTER XXIII.

MENINGITIS AND ENCEPHALITIS.

Inflammation of the coverings of the brain, particularly of the pia (lepto-meningitis), is at times secondary to disease of the brain proper. More often it represents the first and most important factor of a morbid process. The anatomical designation, meningitis, may be applied to a number of different diseases. The symptoms are very much the same in all these forms of disease, and such variation as occurs depends in part upon the intensity of the process, and in part upon the topographical distribution of the disease. It is natural, therefore, to expect that the symptoms of a traumatic or of an idiopathic meningitis, which, as a rule, involves the convexity of the brain, will be different in certain respects from those of a tuberculous meningitis, which involves the base of the brain; not forgetting that the tuberculous process *per se* is responsible for the severity of the symptoms, and the rapid course which that disease runs.

We shall distinguish between—I., Simple acute meningitis; II., Tuberculous meningitis; III., Epidemic cerebro-spinal meningitis, and IV., Meningitis due to various causes.

SIMPLE ACUTE MENINGITIS.

This form of disease occurs after many infectious diseases (pneumonia, ulcerative endocarditis, erysipelas, etc.); after slight or severe injuries, after insolation, and in connection with acute nephritis. The symptoms of acute meningitis are very similar to those observed in all the forms, except that they point to an involvement of the convexity

much more frequently than to an involvement of the base. The disease begins, as a rule, with a feeling of malaise, with vertigo, with nausea and vomiting. These symptoms may remit for a few days or even a week, or they may progress continuously until the headaches become distressing and the vertigo so intense that the child cannot stand, while the nausea and vomiting are repeated at frequent intervals. The vomiting has in most instances the true cerebral character (projectile vomiting). It occurs both after taking food and independently of this. If the vomiting under these circumstances is associated with a clean tongue, its cerebral origin becomes all the more probable. But too much stress should not be laid upon this one point, for gastric disturbance is so common in children, and so often precedes the onset of severe cerebral disease that the presence of a coated tongue need not suggest the improbability of cerebral disease. By degrees the child becomes listless and apathetic, it begins to be drowsy, and sleeps a great deal, and its existence is taken up with sleeping and crying. A convulsive seizure often occurs during this stage. The child becomes irritable, restless, shuns the light (photophobia). The temperature varies between 101° and 104° F.; the pulse is rapid at first, then becomes irregular and slow; the pupils are contracted for a time, later on dilated. The child grows more and more unmanageable until the apathy deepens, and it finally passes into a comatose condition. Before the condition of coma is reached in the vast majority of the cases slight rigidity of the neck sets in, and the upper as well as the lower extremities often exhibit marked spasticity. The deep reflexes are, as a rule, increased. The abdomen is retracted; the bowels are constipated, and if the child is in a deeply comatose condition all efforts at feeding it, or at making it take the breast, are entirely unavailing.

According to the severity of the disease these symptoms will be developed in a shorter or longer period of time, but as a rule a week from the onset is quite sufficient for a full display of the symptoms. To the symptoms noted above we may add the loss of vesical and rectal control, retention of urine, or involuntary micturition and defecation. Vaso-

motor disturbances are marked; tâches cérébrales occur in the disease, but have no diagnostic significance. In some cases ocular palsies (strabismus, ptosis) may be present, and the optic neuritis may give rise to temporary or permanent blindness. A monoplegic or hemiplegic paralysis having all the symptoms of a cerebral palsy may be developed. If the disease takes an unfavorable turn all the symptoms increase in severity, the rigidity of the neck becomes extreme; opisthotonos is developed; the blindness continues; the coma is deepened; respiration becomes irregular, the Cheyne-Stokes type becoming more marked until respiration ceases altogether. If the patient is to pass on to recovery, the symptoms become stationary for a time after the comatose condition has been reached; then the rigidities are lessened, the optic neuritis may recede, and from day to day there are evidences of returning consciousness.

ETIOLOGY.—The occurrence of acute meningitis in association with acute infectious diseases will be referred to again. "Idiopathic meningitis" has been held by many to be a cover for our ignorance. There is no doubt, however, that meningitis may be developed without assignable cause in a child that comes of healthy parents, and that has itself enjoyed perfect health up to the time of the onset of the disease. One variety may be attributed to the effect of intense heat (sunstroke). It is quite likely that a considerable number of the so-called idiopathic forms are due to injuries, the traumatic factor being so slight at times that it does not receive the attention which it merits.

About ten years ago a child was seen in my clinic that had fallen from its crib, a fact which the mother mentioned quite casually. When the child was first examined it presented the typical symptoms of the first stage of a general convexity meningitis. It was slightly stuporous, unable to stand or sit alone, the head was stiff, and it had had occasional spells of vomiting. This stuporous condition soon passed into one of deep coma, the child lost ground rapidly, never exhibiting, however, any cranial-nerve symptoms. After a period of at least six weeks, during which time the child was more or less somnolent, a favorable change set in, consciousness began to return, and the child recovered fully from what was evidently a simple traumatic meningitis.

MORBID ANATOMY AND PATHOLOGY. — Simple acute meningitis is characterized by an inflammatory condition

of the pia mater, which is, as a rule, attended by slight inflammation in the dura and in the gray matter of the brain. In contradistinction to other forms this inflammation is of a serous, or at least of a non-purulent character. There is an increase of cerebro-spinal fluid, the arachnoid may appear a trifle opaque, while the substance of the brain is distinctly œdematous, and even watery. The ventricles are distended, and there may be a condition corresponding to an acute hydrocephalus. The pia of the convexity of both hemispheres is the part most extensively diseased, the pia of the base often being entirely free from all disease, though a slightly increased exudation of lymph may be apparent in the interpeduncular space.

On microscopical examination the blood-vessels of the pia are generally found to be slightly engorged, and an extravasation of white blood-corpuscles is found in the vicinity of the blood-vessels. If the disease has lasted for any considerable length of time, the pia and the cortical substances have become agglutinated so that the pia cannot be removed without tearing the outer layer of gray matter.

Diagnosis.—The diagnosis of simple acute meningitis rests upon the recognition of the symptoms common to all forms of meningitis. These are headache, vomiting, coma, and convulsions; irregularity of the pulse and unequal pupils. It is well to note also the absence of those factors which accompany the graver forms of meningitis (high fever, basilar symptoms, rapid emaciation, and a rapid increase in all the symptoms). The differential diagnosis will not be an easy one, and sometimes a positive diagnosis cannot be reached until the disease takes an unexpectedly favorable turn, or until the patient is seen upon the post-mortem table. But the mistake that is most frequently made is that the meningeal symptoms accompanying many acute infectious diseases are at once pronounced to be the symptoms of true meningitis. This diagnosis has, to the author's knowledge, been made in the first stages of typhoid fever, and in measles and scarlet fever beginning with convulsions; the error is common in otitis and in infantile spinal paralysis that is attended by convulsive seizures and high fever. It is well to remember that though these symptoms resem-

ble those due to meningitis, they are frequently associated with other diseases, and that no physician should be in a hurry to diagnosticate meningitis, pure and simple, unless other diseases can be safely excluded.

COURSE AND PROGNOSIS.—The course of a simple meningitis will almost invariably cover a period varying from four to twelve weeks, or even longer. In those cases that do not tend to recovery the symptoms gradually become intensified, respiration becomes more and more difficult, and after dragging along for some weeks, now and again yielding a hope of recovery, the child finally succumbs from mere exhaustion, from some intercurrent disease, such as bronchitis or pneumonia, or from the effects of cystitis, bedsores, and the like.

In those cases that recover after the coma has lasted for a period varying from one to three or four weeks, signs of returning consciousness are noticed, the child opening its eyes, voluntarily looking around, again taking hold of the bedclothes, of the hand of the nurse or mother, and so on. By degrees the sight improves, hearing, if diminished or increased, becomes normal, the child begins to take its food properly, and from week to week a distinct improvement is noticeable, until all the symptoms have disappeared. But not all of the cases that recover end thus fortunately. The disease does not necessarily kill, but it often leaves distinct traces behind. Not a few of those who are permanently blind owe their misfortune to a meningitis in early years. Permanent paralysis and contractures of one or more extremities are attributable to the same cause, and defective speech, and, above all, a defective intellect are very often the unfortunate outcome of meningitis in early life. Such deformities and defects do not, however, result as frequently from simple meningitis as from the severer types, from which recovery is, on the whole, much rarer.

TREATMENT.—In every form of acute meningitis, whatever its origin may be, I am in favor of adopting the following procedure:

First, keep the patient absolutely quiet, and in a semi-darkened room, and secure the services of a careful and intelligent nurse. Give the child an efficient purgative;

none will do better than calomel, which has attained a certain dignity in all diseases of the central nervous system. Place an ice-bag over the convexity of the skull, or on the nape of the neck if the symptoms point to the involvement of the basilar portion. Though we cannot claim any direct therapeutic effect it will do no harm, for it will at least help a little to reduce the general rise of temperature, and is useful as the first point of attack in treatment. Place the child at once, according to its age, upon moderate doses of the bromide and iodide of sodium (three to five grains of each every four hours). Give these in simple water, or in milk. This treatment may well be persisted in for a few days. If no effect is observed, while it is well to continue the salts of sodium, other more energetic measures may be employed in addition. Among these I would place, first, alternate lukewarm and cold douching of the nape of the neck and the upper portion of the spine, and inunctions of some form of mercury, either of the ten per cent. oleate of mercury or of the unguentum hydrargyri. Inunctions should be performed by the nurse or the mother, and done so thoroughly that the mercury disappears into the skin of the patient.

For purposes of diagnosis, as well as for therapeutic reasons, lumbar puncture may be attempted; in those cases in which the symptoms point to greatly increased intracranial pressure, it will be best to tap the spinal canal on successive or on alternate days, withdrawing only about 10 c.c. at any one time.

After the patient has made a fair recovery, the general condition of the system will need looking after, and a thorough course of cod-liver oil, of malt, or of iron, will be quite in order, and for some months after recovery from this severe disease, the child should be kept free from all excitement and from all mental and physical fatigue. A restful out-of-door life is the very best conclusion to this method of treatment. To patients who exhibit the least tendency to paralysis or contracture, massage should be given as soon as the acute symptoms have disappeared, and electric treatment should be applied to the parts that are distinctly paralyzed.

TUBERCULOUS MENINGITIS

Tuberculous meningitis is by far the most frequent form of meningeal disease in children, if we except the ravages in recent years, and in this community, of the epidemic form of cerebro-spinal meningitis.

The disease itself was first recognized by Robert Whytt, in 1768, and since that time has been the subject of innumerable articles; but every point regarding the disease is now so thoroughly known that we can sum up its chief characteristics without referring to individual authors. Since Koch's discovery of the bacillus of tuberculosis there has been no doubt of the microbic origin of tuberculous meningitis. According to Holt's statistics, twenty-five per cent of all deaths from tuberculosis in children were due to meningitis. This form of meningitis is always secondary.

The disease occurs in children much more frequently than in adults; while it is rare in young infants, it is more frequent in children between the ages of six months and four years. In families in which there is a distinct hereditary taint of tuberculosis or scrofula, children that have been apparently well are suddenly attacked by this malignant disease, and healthy children in families in which every hereditary predisposition is wanting, or, at least, denied, are also affected. But in the majority of instances the children attacked have been weak and feeble; many of them have suffered from chronic intestinal troubles, from swellings of the glands, from nasal and aural catarrhs, in short, from those conditions from which we may safely argue a tubercular infection. A chronic laryngitis or bronchitis, or a swelling of the bronchial glands, which may not have given rise to any special symptoms, is often the precursor of tuberculous meningitis.

ONSET.—In contradistinction to other forms of meningitis, that dependent upon tubercular infection comes on in a very insidious fashion. The child first loses its brightness and cheerfulness, and complains of an occasional headache, and slight nausea; a vomiting spell may occur. The pulse is generally rapid. These complaints almost invariably lead to the suspicion of slight gastric disturbance, which, fortunately, is quite often the case; but every physician will

do well, whenever this series of phenomena occurs, to be watchful, and to give notice that he should be informed if the symptoms do not promptly disappear. There may be a marked remission or great improvement for a few days; then a change takes place, the nausea becomes more frequent, the headache more intense, and the vomiting spells are a daily occurrence. In this way a week or more may pass, the physician and parents hoping that all the symptoms may subside. At the end of about a week little doubt remains of the significance of the condition, for if a child is afflicted with a tuberculous meningitis the headaches become intense and persistent, the child gives the short hydrocephalic cry, and by degrees becomes somnolent; slight rigidity of the neck is observed, the pulse has become slower, and the child shows every sign of serious illness. Aphasia may be an early symptom; and all signs point to an intense general brain disturbance, and to the localization of the disease chiefly at the base of the brain, from which the various cranial nerves issue.

The temperature is subject to great variations, and during the first few weeks it does not, as a rule, rise above 103° F., but during the last week it may reach 105° F., and during the terminal stages may reach 106° F., or even higher. In one case, an hour before death, I recorded a temperature of 107° F. This is supposed to be due to a paralysis of the heat-regulating centres. Respiration is not seriously interfered with, as a rule, until the child enters upon the terminal period, during which stage the breathing becomes irregular, often of the typical Cheynes-Stokes type, and cyanosis is added to the host of other symptoms.

As we examine the child from head to foot, we are apt to find a variety of symptoms. Rigidity of the neck, with or without opisthotonos, and excessive painfulness on every passive movement of the head or trunk. The majority of the patients present distinct convergent strabismus, due, as a rule, to a paralysis of one or more of the ocular muscles. The pupils are unequal and dilated, contracting very sluggishly to light, and their reaction during accommodation can, of course, not be tested in consequence of the comatose condition. The conjunctival reflex is lost at an early period, and in consequence of diminished movement

of the eyelids the cornea becomes cloudy easily. Deglutition can be carried on only imperfectly, and during an effort to open the mouth or to perform chewing motions, a trismus is very apt to set in. A further examination of the head may reveal a paralysis of some of the branches of the facial, the paresis of these muscles being, at times, unilateral and at other times bilateral; but the paresis is evidently due to involvement of the nerves at the base, and if an examination is made to bring out this special point, the electrical reactions may be found altered in keeping with this special localization of the lesion.

An examination of the eyes will disclose a hyperæmic and swollen condition of the papillæ. In some instances there may be a typical optic neuritis, single or double.*

The abdomen is retracted, tâches cérébrales are easily produced (not pathognomonic).

The upper and lower extremities may be paralyzed to a greater or lesser degree; the exact amount of paralysis cannot always be easily determined on account of the entire absence of voluntary action, and because the physician is unable to make satisfactory tests for this special point. While the paralysis may be of sufficient interest as illustrating the exact distribution of the meningeal process it is of very little practical importance, and loses in value as compared with the other and more serious symptoms. The paralysis may be hemiplegic; in some forms it is bilateral or irregular. The cutaneous reflexes are diminished, often

*Much has been made of the presence of tubercles in the choroid, and it has been claimed by many that the existence of these tubercles is one of the most important symptoms of the earlier stages of the disease. The truth of this cannot be gainsaid, but as a matter of practical experience it must be admitted that in many cases of undoubted tubercular meningitis the expert oculist does not find tubercles, and that in those very cases in which the discovery of such a tubercle would have helped to determine the exact character of a meningitis, the tubercles cannot be seen, although later post-mortem examination leaves no doubt of the tubercular character of the process. While we may regard the presence of tubercles, therefore, as a valuable corroborative symptom, a failure to detect them should not be allowed to disprove the diagnosis if other symptoms would seem to point to the tubercular nature of the trouble. Dr. R. L. Randolph has examined thirty-five cases of meningitis of all kinds with the ophthalmoscope, and found the fundus normal in seven patients. The optic disks were usually congested, with the retinal vessels distended and remarkably tortuous In three cases the entire eye was normal, and these three patients recovered. In the four fatal cases with normal fundus some other ocular symptoms were present.

lost. The deep reflexes in the upper and lower extremities are, as a rule, increased. Kernig's symptom is generally present.

Epileptiform convulsions are not rare, both at the onset of the disease and during later stages. These convulsions are, as a rule, general in distribution, and not of the Jacksonian type. If the latter form should prevail, there would be good reason to infer that the cortex is diseased as well as the base of the brain. Considering the fact of the existence of so-called epileptiform centres in the pons and medulla, the wonder is not that such convulsions occur, but that they do not occur much more frequently. In the terminal stages of the disease convulsions become more frequent, as they do in many other brain diseases; and in tubercular meningitis the frequent occurrence of convulsions, with a rise in temperature, may be taken to be the sign of the approaching end. In the terminal period, too, the paralysis becomes complete, the pupils are dilated, the tongue dry and furred and the temperature may fall to 93° or 94° F., until an ante-mortem elevation of temperature begins.

All the phenomena are remarkably persistent during the course of this disease. There are at times slight remissions in the ocular palsies, but after the symptoms have once been fully developed, they remain very much the same to the end. The changes which take place are these: Respiration becomes irregular, at times intermittent, the pulse grows feebler and slower, deglutition more and more difficult, and the child dies from paralysis of the cardiac and respiratory centres.

Morbid Anatomy and Pathology.—Every one who has had an opportunity to remove the brain from cases of tuberculous meningitis is surprised by the few changes found in this organ. In the great majority of autopsies, on the removal of the calvarium there is very little evidence of any active process on the convexity. In some brains the pial vessels are much congested, the sinuses are filled with clots that have evidently been formed only a few days before death, and the hemispheres in general present a more or less œdematous appearance.

Minute tubercles are noticed along the distribution of some of the larger pial veins on the convexity. The chief

changes cannot be noted until the brain has been removed from the skull. At the base of the brain the character of the disease is easily recognized, the pia is cloudy, and in some places bulging because of the accumulation of fluid underneath. This is particularly noticeable in the interpeduncular spaces, in which the tubercles are, as a rule, most freely developed. These tubercles, often no larger than a pin's head, may be scattered throughout the entire pia from the optic chiasm to the pons, medulla, and spinal cord. The presence of tubercles is not always established at the time of the autopsy, but this need not militate against the proper diagnosis, for it is a fact, well proven, that in undoubted tubercular diseases we may have an involvement of the pia without tubercles, and occasionally the presence of tubercles with but few signs of an inflammatory process.

PATHOLOGY.—Of the pathology of tubercular meningitis little need be said, as it belongs to the order of infectious disorders, and the disease is, in almost every instance, a part of a general tubercular infection. The tubercular form is, moreover, rarely a primary affection, although it sometimes occurs in children who have been apparently healthy. A thorough post-mortem examination reveals quite regularly distinct foci of disease in the lungs, in the mesenteric glands from which the infection in all probability took its start. The invariably fatal issue must be attributed in part to the general effect of the tubercular poison and its ravages in other organs; for the remarkably slight changes in some of the cases that have taken a rapid course are the surprise of every pathologist. A few small tubercles, without much exudation, even though they be in the vicinity of the pons and medulla, can hardly be considered a sufficient cause of death, whereas the toxines circulating in the blood may have been the initial cause of the paralysis of the vital centres.

DIAGNOSIS.—The difficulties of a differential diagnosis* are not limited merely to the period of onset, during which

* The general practitioner is apt to attach too much importance to the general symptoms, and too little to the local (basilar) symptoms. The pulse, the temperature, the condition of the abdomen, may leave the diagnosis in doubt, but an acute ocular palsy, however slight, or an incipient optic neuritis, will indicate the true nature of the trouble.

time the most experienced physician may well be in doubt as to the true character of the disease. But the more difficult question arises later on, whether the disease be a simple meningitis, whether it be of tubercular origin, or whether it represents an epidemic form. In order to determine this, it is best to keep in mind the antecedent history of the child; a history of tubercular trouble in the family, or of an early scrofulous or chronic catarrhal condition would naturally prejudice the physician in favor of a diagnosis of tubercular trouble, but everyone has experienced curious surprises in this respect, for children of tubercular ancestry may have a simple meningitis which is recoverable, or they may be afflicted with the epidemic form. The diagnosis will be confirmed by the presence of tubercle bacilli in the spinal fluid, or according to French authors, by the presence of mononuclear lymphoid cells. The absence of bacilli need not militate against the diagnosis. In addition, it is fairly safe to infer that cases of tubercular meningitis invariably present basilar symptoms, whereas the symptoms pointing to involvement of the convexity only, occur more frequently in the other forms.

Course and Prognosis.—Tuberculois meningitis runs, as a rule, a course varying between three and six weeks, though the time may be extended a little if the premonitory period is taken into account. In the more virulent forms death may occur at the end of the first week, or in the course of ten or twelve days; and in these forms I have often found only slight post-mortem changes, from which we may infer that the general toxine poisoning has been of much more consequence than the local deposit at the base of the brain. As for the prognosis, no one need hesitate to say that it is absolutely bad; but in making such a prognosis the most experienced physician will do well to remember that this diagnosis is never quite as certain as is the prognosis based upon it. There is grave doubt whether cases of tuberculous meningitis ever recover.

Henoch, and Rilliet and Barthez, record two cases in which death ensued from a second attack, occurring some years after the first; but even here there is room for doubt as to whether the first attack of meningitis was truly tuber-

cular in character. Pollitzer reports a case of a child which survived three years after an attack of basilar meningitis; at the autopsy he found, at the base, the distinct evidences of an old exudation over the pons, which was in all probability of a tuberculous nature. Freyhan found the tubercle bacilli in the cerebro-spinal fluid of a patient who recovered.

Oppenheim has reported six cases of a localized tuberculous meningo-encephalitis of the Rolandic areas in children, simulating brain tumor, ending in recovery. The anatomical proof is wanting, but the author's hypothesis is worth considering.

TREATMENT.—The patient should be put as quickly as possible in a quiet, darkened room. Administer calomel at once, in a sufficient dose to effect a very copious discharge from the bowels. If the child will tolerate it, put an ice-bag over the nape of the neck; or, if the ice-bag is unpleasant, ordinary cold applications can be tried. As soon as the bowels have been moved give the iodides, the bromides, or mercury; employ those therapeutic measures which we have discussed in connection with other forms of meningitis.

From the very beginning, too, observe the cardiac and respiratory functions, and give mild cardiac stimulants. The best are small doses of digitalis and caffeine.

Special attention should be paid to the feeding of the child. Feeding by the spoon and giving the food in very small quantities at a time is the only proper method. If great care is not exercised the liquids may flow into the trachea and produce very uncomfortable symptoms, with the possible result of complicating pneumonia. According to the condition of the child the physician should exercise his judgment, and remember that loss of sleep and lack of quiet are often much more harmful than lack of food, and every child suffering from any form of meningitis should be given ample time for quiet sleep. The old habit of insisting on half-hourly, or even hourly, feeding is not to be commended in these cases.

Unfortunately there is little reason with the majority of patients to change this method of treatment, for whatever method be employed the results are equally disastrous; but a persistent effort should be made, and the attempt to conquer the disease should not be given up until the child begins to decline rapidly, until it fails to swallow food, for

from that time on medication will do very little good. Persistent administration of medicines, of nutrient enemata, and the like, beneficial as they may be in other diseases, in these simply tend to prolong the agony of the child, and of the careworn parents or relatives. Lumbar puncture is of no avail in this form of meningitis, and, after the diagnosis has been established, should not be repeated.

Surgical interference has been attempted in tubercular meningitis. Ord and Waterhouse have trephined a case diagnosticated as tubercular meningitis, and have drained the subarachnoid space;* the child recovered. There is reason to doubt the tubercular character of the disease in this case, but the relief after the operation was so marked that the propriety of an operation can be entertained before the child is exhausted, if pressure symptoms are extreme and the character of the meningitis is doubtful. A recent author (Hirschberg) believes that death is due, not to the tubercles, but to intracranial compression. We have stated on a preceding page that the increase in intracranial fluid is often very slight indeed. The trephine in Ord's patient was applied midway between the external occipital crest and mastoid process. The operation has been attempted in this city on an undoubted case of tubercular meningitis and the child died very shortly thereafter.

EPIDEMIC CEREBRO-SPINAL MENINGITIS.†

This special form of meningitis has attained a sad distinction in many countries. In America its ravages have been very much greater than in Europe.‡ In New York City serious epidemics occurred in the winter and spring of 1904 and 1905. It belongs to the category of infectious fevers. The microbic origin of the disease has recently been established beyond peradventure, though it seems to be still a matter of doubt as to whether one or more microorganisms bear an etiological relation to it.

* Such drainage could be effected quite as readily by a lumbar puncture according to Quincke's method.

† Synonyms: "Spotted fever," "cerebro-spinal fever," "typhus petechialis," "febris nigra," and "malignant meningitis," are the common synonyms, all of them indicating the infectious and grave character of the disease

‡ At this present time (1905) serious epidemics of "spotted fever" have been reported from various parts of Continental Europe. In eastern Prussia the disease appeared in its most virulent form; and if the earliest statistics are reliable, the mortality rate is about the same as in American cities. In spite of "commissions" that have been appointed, no sovereign remedy has been discovered.

It is the accepted opinion of the day that this grave disease is due to infection by the diplococcus intracellularis (Weichselbaum, Jaeger, Heubner, Osler, Koplik, Holt, and others). Some sporadic cases have been shown to be due to the micrococcus pneumoniæ of Fraenkel, and in some cases, resembling the epidemic form clinically, streptococci and other micro-organisms have been found. The only just inference is, that various organisms may produce cerebro-spinal meningitis, but that the epidemic form, and no doubt many of the sporadic cases, are due to the diplococcus intracellularis.

The epidemics have varied much in severity, and in each epidemic there have been some mild, and an alarming number of most virulent cases. The micro-organisms have been found in the nasal secretions, thus suggesting a possible point of entrance into the cranial cavity. Among the New York physicians reporting upon the latest epidemics are Chapin, Nammack, Manges, Koplik, Lambert, Berg, and others.

The first epidemic of cerebro-spinal meningitis was distinctly recognized as such, and well described under the heading of "a malignant non-contagious fever" by Vieussens in 1805, who described the disease as it appeared in Geneva. Thirty-three persons lost their lives during this epidemic; the average duration of the disease was from one-half to five days. A few years later epidemics occurred in various parts of Germany, in Holland, and in England. Dr. J. Lewis Smith, to whom we are indebted for one of the best contributions to our knowledge of this disease, states that the first American case occurred at Medfield, Mass., in 1806. From 1806 to 1816 it appeared in various localities both in Canada and in the United States. Between 1816 and 1828 one epidemic occurred at Middletown, Conn., and another at Vesoul, in France. In 1833 Naples was visited by this epidemic, and the disease did not appear again until 1837, and then various localities in France were stricken. The military were chiefly affected by the disease, and a very large proportion of those affected died from it. Between 1837 and 1849 France was the chief seat of the epidemic. In the next ten years almost every part of Europe was visited by the disease. In 1842 another epidemic broke out in the United States, at a distance from the sea-coast, and as Dr. Smith says, apparently not by communication from Europe; but this could hardly be maintained with our present views regarding the transmission of micro-organisms from one country to another. Epidemics occurred in States as widely apart as Alabama and Mississippi, New York and Louisiana. Norway and Sweden were the chief seats of the disease between 1854 and 1860, and since that time scarcely a single city or district has been entirely free from the disease. Inasmuch as isolated districts have been affected, the disease was not supposed to spread in the manner of ordinary contagious diseases, but to have been engendered by local conditions, among which the massing together of large classes of population in poorly ventilated and filthy quarters, as in military barracks for instance, was considered to be the most favorable predisposing cause. The disease is now permanently established in almost every large American city, though it has rarely assumed a severe epidemic form. In New York City, from 1866 to 1872, the

annual deaths from this disease, according to Dr. Smith's statistics, varied from eighteen to forty-eight. A very severe epidemic occurred in December, 1871, and lasted well into the summer of 1872, so that 782 deaths, chiefly in children, resulted from cerebro-spinal fever in this city alone. Since that time the annual deaths have varied between 97, in 1878, and 461, in 1881. An important epidemic, though a small one, was the epidemic of Lonaconing, which was most carefully observed by two competent physicians, of which an account is given in the article by Flexner and Barker in the year 1894. This town is situated in the Alleghany Mountains and contains about five thousand inhabitants. A muddy stream which passes through the town, receiving most of the sewage, appears to have been responsible for the spreading of the epidemic. All the conditions, including the overcrowding of miners in filthy houses, were favorable to the spread of the disease.

The disease is common both to man and beast. A serious epidemic occurred in New York City in 1871, and was at once recognized as a filth disease, for it first affected the horses in the large and overcrowded stables of the car and stage lines. A few individuals were soon similarly afflicted, but it is doubtful whether the disease was transmitted from the animals to the men who were in charge of them. The epidemic which occurred in 1872 was evidently related to this same outbreak in animals in 1871. Though the disease is bred by filth, it may unquestionably also be carried, or at least transmitted, to persons, and particularly to children living in excellent hygienic surroundings, and many of us have seen such cases in the habitations of the richest as well as in the families of the working-classes. The disease may attack those in good health, but is even more apt to strike those whose health has been injured by previous disease or by fatigue. Dr. Smith quotes Frothingham as an authority for the statement that in a brigade of the Army of the Potomac which was attacked by this epidemic, the men were almost exhausted from excessive drilling.

The disease shows no distinction between the sexes, at least in children. Dr. Smith reports 105 cases occurring in his practice, of which 59 were in males and 46 in females; 91 of these cases being in children. While persons of every age may be attacked by the disease, it is unquestionably more liable to attack children in the earlier years of life than at any other period. It is interesting to note the statistics for a single year, 1883, as given by Smith for New York City:

Under 1 year	57	From 15 to 20 years	15
From 1 to 2 years	31	From 20 to 25 "	7
From 2 to 3 "	22	From 25 to 30 "	3
From 3 to 4 "	12	From 30 to 35 "	4
From 4 to 5 "	9	From 35 to 40 "	3
From 5 to 10 "	37	After this scattering cases.	
From 10 to 15 "	18		

The New York Health Department records 498 deaths from cerebro-spinal meningitis from March 12th to May 21st, 1904.

SYMPTOMS.—The majority of the symptoms will naturally resemble those occurring in other forms of meningitis. The onset of the disease is characteristically sudden. A child in perfect health is suddenly seized with headache, vomiting, and either slight rigor or convulsions. In the milder cases a few days of malaise and of slight nausea may precede the onset of the other symptoms. The first severe symptoms are promptly followed by a stupor, which is apt soon to deepen into profound coma. I remember the case of a child, one year of age, in which a slight headache, vomiting, convulsions, rigidity of the neck, deep coma, and strabismus all developed within twenty-four hours, and death ensued within forty-eight. Severe neuralgic pains are frequent in the earlier stages of the disease. The pupils are unequal and dilated. The vomiting is of the cerebral order and occurs on the first or the second day in the vast majority of the cases. The fever is, as a rule, high, varying between 103° and 105° F.; in one rapidly fatal case it reached 107° F. a few hours before death on the second day.

The meningeal symptoms are developed in a very much shorter period of time than in other forms of meningitis. We do not have the slow progress from stupor to coma which we find in the tubercular type; often the coma is deep from the very beginning. Delirium may alternate with intense coma. In some of the cases in which the coma is not profound, great restlessness takes the place of the stuporous condition. The child is, as a rule, extremely sensitive, even hyperæsthetic. The slightest touch of any part of the body, the mere weight of the bedclothes, is often sufficient to elicit shrill cries. This hyperæsthesia is explained quite readily by the irritation of all the posterior root-fibres by the meningeal exudate. Contractions of the various muscles occur at an early stage of the disease. The head is firmly retracted, opisthotonos is distinctly developed, the thighs and legs are in a flexed position, and the arms and hands may be distinctly contractured. A hemiplegia or an alternate form of paralysis, or a monoplegia, together with cranial nerve palsies, may be made out in some patients. The deep reflexes are exaggerated, and the superficial

reflexes are diminished. Kernig's sign is present. Convulsions are very common.

The author saw during the spring months of 1904 a number of fulminant cases in which the disease ran its entire course within forty-eight hours. In such cases the chief symptoms are: Headaches, high fever, rapidly developing delirium, rigidity of the neck; in short, general, not localizing, symptoms.

The "tâche cérébrale" was formerly considered to be a rather important symptom of all forms of meningitis; but it has lost its pathognomonic value, as it occurs in many other diseases. The skin presents a peculiar mottling in the first or second week of the disease, and particularly when the temperature is low. Small red points and large bluish spots, due to exudation of blood under the surface, also appear, and were seen frequently enough to justify the term "spotted fever." In European epidemics these peculiar extravasations have not been regularly observed. Herpes occurs, and Smith refers to the occurrence of erysipelas; but the latter is evidently entirely independent of the meningitis; it is a complicating condition, and not in any sense a symptom of the epidemic form.

The organs of special senses are frequently affected in epidemic meningitis, and possibly more frequently than in other forms. A hyperæmic and inflammatory condition of the entire eyeball is a common occurrence. The media may become cloudy and the various structures may become adherent to one another; occasionally ulcerations of the cornea and perforation of the eye with total loss of vision may occur. According to Knapp, as quoted by Smith, the nature of the eye affection is a purulent choroiditis, probably metastatic. In some cases a double optic neuritis occurs, and from this, as well as from the inflammatory conditions of the eyeball, total blindness may result. Not a few of those who recover from cerebro-spinal meningitis are afflicted with permanent blindness as the result of this dreadful scourge.

The hearing is often as seriously impaired as vision, and severe otitis media, ending in suppuration, with perforation of the drum and all its sequences, is a common occurrence. In other instances loss of hearing is evidently due to more central causes, and is developed only after recovery from the main disease. It is unfortunate that the loss of hearing is apt to be bilateral and complete. According to Smith's statistics from the epidemic in 1872, about one in every ten patients became deaf. But he states that in the milder form of cerebro-spinal meningitis which has prevailed since 1872 the proportionate number that has been thus affected has been less, and the same may be said with reference to loss of sight. Knapp reports that

among twenty-nine cases of total deafness occurring after cerebro-spinal meningitis only one seemed to give evidence of hearing afterward.

Morbid Anatomy and Pathology.—We need not again insist upon the microbic origin of the disease. If the exudate is examined the diplococcus intracellularis or the pneumococcus may be found. The chief anatomical characteristics of the disease are an intense hyperæmic condition of the meninges and of the brain, and this may be the sole morbid condition if the patient has died in the very early stage of the disease. If it has lasted more than a few days, pus is visible to the naked eye under the arachnoid. This membrane loses its transparency and begins to appear cloudy, the cloudiness being most apparent along the course of the vessels from which the exudation undoubtedly takes place.

The pus is found both in the meshes of the pia, and under the pia, between it and the cortex.* The fibrinous purulent layer will be found adherent to the pia, and can usually be removed together with this membrane. These purulent layers can be drawn out of the fissures, leaving discolored tissue underneath. This purulent exudation covers not only the fissures of the convexity, but extends with equal frequency over the base of the brain, and in those very spaces in which we are accustomed to look for tubercular deposits. In addition to the exudation over the brain proper, an equally thick layer can often be found over the greater part of the spinal cord, holding the same relations to the spinal meninges that it does to the cerebral coverings. The blood is apt to be clotted in the large veins and sinuses, and such clots may be of a purulent character. The ventricular fluid is, as a rule, increased, and in the more violent forms may contain small floccules of fibrin or fibrinous pus.

In addition we may find hypostatic pneumonia, or varying degrees of bronchitis and atelectasis; all the serous membranes may be in a condition of inflammation. The spleen is almost invariably enlarged, while the other abdominal viscera are found in a condition of decided hyperæmia; the kidneys may be in a condition of acute congestion, and, according to Welch, an acute diffuse nephritis is occasionally present.

* An excellent colored plate of a purulent meningitis may be found in Holt's textbook, page 752.

Differential Diagnosis.—The only point to be considered in this respect is the differentiation of this form from other forms of meningitis. This differentiation should not depend solely upon the bacteriologic findings, but note that the disease advances much more rapidly than do the tubercular or other types. The mental symptoms are, above all things, developed much more actively and vehemently, in keeping with the early exudation of pus over the surface of the brain. The temperature is higher in the earliest period of the disease. Cranial nerve palsies are often wanting. The occurrence of similar disease in the city, the condition of the environment of the patient, and the exclusion of those facts which tend to prove the presence of tubercular disease will help to make the diagnosis of the epidemic form much more certain.

Prognosis.—The disease is unquestionably one of the most fatal diseases of childhood, and if it does not prove fatal the condition of the survivor is often so distressing that death would have been preferable. About one-half of the patients make a fair recovery. The duration of the coma is as reliable a sign as any in giving a prognosis. Cases in which the coma is rapidly developed and does not show any sign of receding within the first week or two are almost certain to end fatally. If the coma has been slow to develop, the inference is justified that the process is a less intense one, and there is in so far a hope of recovery. But even in these cases if the coma which has once been developed remains stationary for a week or more, the chances of recovery lessen with almost every hour that the coma continues. In spite of all rules that may be laid down children whom we have reason to expect to recover take a turn for the worse, and not a few of those who have been given up by the most careful and experienced physicians make good recoveries; but in others, in whom the disease drags along, the final issue is simply deferred, and death may result from exhaustion, as in one of my own cases, as late as four months after the onset of the disease.

Treatment.—The treatment of this special form of meningitis can differ in no respect from that noted in connection with the discussion of the tubercular and simple

forms of meningitis. Although the disease has not been proved to be contagious, the patient should be strictly isolated.

In the epidemic of 1904, Manges, following the recommendation of Seager, who reported upon an epidemic in Lisbon, advocated the use of a three to nine c. c. injection of a one per cent. Lysol solution into the spinal canal after the withdrawal of cerebro-spinal fluid. Three cases, treated in this fashion (one of them a streptococcus infection) recovered. In the hands of others this treatment has failed. During the present epidemic, the suggestion has been made to use the Diphtheria Antitoxin in the treatment of epidemic cerebro-spinal meningitis. If such treatment prove successful, the Bacteriologists will have to revise their fundamental doctrines and beliefs.

MENINGITIS DUE TO OTHER CAUSES.*

I. Meningitis due to traumatism has been mentioned in connection with acute meningitis; it is much rarer in children than in adults, for reasons which it is hardly necessary to explain; though the injuries which children receive may be relatively slight, meningeal disturbances follow upon them in some instances. A fall from a chair, a fall down the stairs, or a blow dealt upon the head of the child, to which little heed is paid at the time, may be the starting-point of the meningitis. At times the same slight traumatic factor is the cause of an epilepsy developing after six months, or even after a year, or still later (see chapter on Epilepsy). In these children a local meningitis or meningo-encephalitis may be considered to be the actual lesion responsible for the epilepsy. With such cases we have no concern at present. We must take into account, however, those patients in whom some traumatism to the skull is followed by the symptoms characteristic of meningitis. The inflammatory process may involve both the dura and the pia, and may be either serous or purulent.

In these days of cranial surgery, another form of (purulent) meningitis is to be observed in children and in adults after operative procedures for the relief of epilepsy or other brain diseases. The aseptic principles of modern surgery are calculated to prevent such complications. It is exceptional to see this condition in cases in which a simple trephining operation has been done, but if the dura has been opened the danger of meningitis is greater.

* For "Meningitis serosa" see page 421.

The prognosis of a meningitis developing after operation is, on the whole, excessively grave. The only advice to be given is that as soon as the temperature rises, and as soon as the first symptoms of a suppurative meningitis set in, the wound should be opened at once, and an effort made to treat the condition according to the best surgical principles. The same applies to those cases of meningitis which follow upon external injuries, if the meningeal symptoms can be clearly traced to the preceding injury. A careful examination of the skull should be made in order to determine, if possible, where the injury has been inflicted. If the slightest depression can be felt, trephining should be done over this region, and if no changes in the skull can be made out, but the symptoms point to an incipient meningitis, any abrasion of the scalp, or any extravasation of blood under the scalp, should be a sufficient guide as to the site of the operation. I am firmly convinced that much good can be done by proper and timely surgical interference in these cases, and little harm will result even if the operation should prove that no tangible local injury has been done to the brain or skull. To be sure the operation should not be undertaken if the child's general condition is such that it will not bear the shock of the operation. In such circumstances the calm judgment of an experienced surgeon or physician will be of great value, but it should be distinctly stated that coma, however profound, or recurrent convulsions, do not constitute a contra-indication to operative interference.*

II. Meningitis in a purulent form may result from disease of the ear. The aurists are well aware of the danger lurking in every form of disease of the mastoid and of the inner or middle ear; for if the pus that is formed in any one of these regions is not discharged outward, the danger of its causing a purulent meningitis by a direct discharge inward, or through caries of the petrous bone, is much to be feared. Chronic ear trouble, so slight that little attention is paid to it, may persist for years before causing a purulent men-

* Too much valuable time is wasted with the administration of drugs that may ease the physician's conscience but do the patient little or no good. If the symptoms point to an increase of intracranial pressure, or to an extension of the morbid process, an exploratory trephining should be attempted.

ingitis.* Since aural surgeons have become accustomed to operate promptly upon the appearance of symptoms pointing to suppuration in the mastoid process, or in the other bony parts of the ear, these forms of purulent meningitis from ear disease are far less frequent than they were formerly. The symptoms which point to an incipient meningitis can be distinguished in many cases, though not in all, from those due to the presence of pus within the ear structures alone. In addition to the intense pain, the presence of pus in the ear may cause giddiness, intense vertigo, vomiting, and even slight stupor, but if the headaches become general and most intense, if the child becomes comatose, if the pulse is either slowed up or very much accelerated, if the neck becomes sensitive and rigid, and if a slight optic neuritis should set in in one or both eyes, it is certain that the pus has passed beyond the limits of the ear and has set up a meningitis, or possibly an abscess. Under such conditions it is imperative to operate, giving the pus a chance to discharge outward, and to lay bare, if necessary, the parts of the brain which are most apt to be involved in ear disease. It will be advisable at the start thoroughly to explore the mastoid, as well as the middle and inner ear, before attempting to trephine over the temporal convolutions, but if the operations upon the bony parts of the ear do not give prompt relief the cranial operation should be done. The aural surgeons are opposed to operations in cases of purulent meningitis, but as long as the meningitis is strictly localized and confined to the parts in immediate juxtaposition to the ear, there is every reason to advise an exploratory operation to secure an exit for the pus, which, if confined to these parts, is bound to cause a general suppurative meningitis.

If suppurative meningitis is due to disease of the mastoid, the pus is more likely to find its way either to the cerebellum or to the base of the brain. In diseases of the middle and inner ear, the meningitis is more apt to be developed

* The author saw this well illustrated in a lad of fourteen (a patient of Dr. Wiener), who since his fourth year had had chronic ear disease; without special cause this old trouble was lighted up and ended in death within one week. The autopsy disclosed a wide-spread purulent meningitis from caries of the right petrous bone. The morbid condition was most marked over the left temporo-sphenoidal lobe.

in the regions over the first and second temporal convolutions. In endeavoring to locate the proper region for opening the skull in cases of ear disease it is best to select a point that is reached by going one and a quarter inches back of the external auditory meatus, and from this point one and a quarter inches upward. If the surgeon remembers that a line drawn from the outer angle of the orbit horizontally across the skull gives the approximate position of the fissure of Sylvius, and therefore the upper limits of the temporal convolutions, he cannot well fail to open the skull over the part of the brain that comes into question in these cases.*

Meningitis may also be caused by a tumor or an abscess in adjacent parts of the brain; but in such cases the meningitis is of relatively little importance, and if it gives rise to distinct symptoms simply helps to intensify those caused by the chief morbid process. Purulent meningitis is sometimes due to disease of the nose, to erysipelas, and to purulent disease of the eye. If so, the antecedent conditions will leave little doubt of the cause of the meningitis, and the subsequent symptoms will not vary from the meningitis following upon other causes.

III. Many of the infectious diseases of children lead to meningitis. Among these are measles, scarlatina, small-pox, and even rheumatism and influenza, but, above all, typhoid fever and pneumonia.

Meningitis has been found present in the autopsies upon persons dead from typhoid fever, but is by no means present in all those forms of typhoid fever in which at the beginning or in the later stage of the disease delirium and coma are developed in connection with the fever. If a child have typhoid, the diagnosis of additional meningitis should not be made unless there are very positive symptoms, such as decided rigidity of the neck, hyperæsthesia of part or of the entire body, and cranial-nerve palsies.

Brain symptoms are not infrequent complications in acute pneumonia, and much attention has been directed to these cerebral complications by Holt and others; but meningeal disease unquestionably occurs in a number of instances, and we can the more readily understand the connection

* See chapter on "Abscess."

between the two diseases since Fraenkel's pneumococcus has been found in the inflamed lung, and in the meningeal exudate. The same coccus is also found at times in the epidemic purulent form of meningitis.

IV. Among the many curious and surprising sequelæ of influenza which have been described, none has been more striking than the occasional occurrence of cerebral and cerebro-spinal meningitis during or immediately after the acute stage of the disease. For the present the evidence is in favor of an encephalitis, rather than a meningitis, as the cause of this special form of disease. (See p. 416.)

Inflammation of the meninges also follows upon general septicæmic processes, but this special causation is much more common in the adult than in the child. The typical septicæmic meningitis is that form which we encounter in connection with the puerperal state in women, and from which I have seen one most remarkable recovery. There is no form of septicæmic meningitis in children quite as typical as this puerperal meningitis, unless it be that which occasionally occurs with septic bone disease or with ulcerative endocarditis.

CHRONIC BASILAR MENINGITIS.

A special form of meningitis occurring in children has been described by Gee, Barlow, Still, and others. Its only claim to recognition is that it may run a protracted course varying from months to a year or more, that it affects very young children, that the inflammation is most marked in the posterior fossa of the base—and that for a long time cervical opisthotonus is the chief symptom. Since Still has shown that the diplococcus intracellularis was found in seven of eight cases, and since these cases have been observed during epidemics of cerebro-spinal meningitis (Koplik), there is no reason to regard chronic basilar meningitis as a distinct clinical entity. The designation "posterior basic meningitis" is misleading; tuberculous forms are "posterior basic" often enough, as well as anterior basic, or middle basic. Such terms produce confusion.

In the cases so classified, lymph may glue together the medulla and the cerebellum, the openings from the fourth ventricle may be closed, and the ventricles distended. The writers advocating this form claim that the symptoms are: Gradual or sudden onset, convulsions, slight or no fever, opisthotonus, general muscular rigidity, mental dulness and irritability, rarely coma. We agree with Holt that some forms of "chronic basilar meningitis" in children may be syphilitic. As for treatment, we would urge lumbar puncture, to be repeated carefully a number of times, and the administration of iodides, about ten or fifteen grains per day, or more—according to the age of the child.

ACUTE HEMORRHAGIC ENCEPHALITIS.

We cannot close the discussion of meningitis without reference to acute encephalitis, which is frequently associated with inflammation of the pia, but is also at times developed independently of any meningeal trouble. In meningitis due to traumatism, or to acute and chronic intoxication, some encephalitis occurs by simple extension of the inflammation. Strümpell, Fürbringer, Leichtenstern, Oppenheim, and others have endeavored to establish a form of acute hemorrhagic encephalitis which is developed commonly after some acute infectious disease, particularly after influenza.

Simple acute encephalitis can be recognized by the following signs: The disease is very apt to attack children before the age of puberty. For a few days previous to the full development of the disease the child complains of headache, of dizziness, and is irritable or depressed; drowsiness, gradually developing into coma, soon sets in. Rigors and slight elevations of temperature point to the development of an acute infectious trouble. The loss of consciousness need not be complete, and distinct remissions occur during the first week of the disease. Hours of stupor are followed by a condition of wakefulness and restlessness. The pupillary reflexes may remain normal or may be sluggish; the deep and superficial reflexes are not altered. A rigidity of the neck and slight opisthotonos are early symptoms. Paralysis, in the form of monoplegia or a hemiplegia, is observed at an early day, and may or may not be associated with aphasia.* Ocular and other cranial nerve palsies are developed, and if they set in in an acute fashion the condition may remind one very easily of a basilar meningitis, with the exception of the incomplete loss of consciousness. Respiration may become irregular, and the cardiac action is either accelerated, irregular, or diminished. Optic neuritis has been observed in several such cases. The symptoms will vary very much according to the seat of the disease. Loss of consciousness, convulsions, and palsies will be more frequent in encephalitis of the convexity, whereas cranial nerve palsies, dizziness, difficulties in deglutition and in articulation may be expected if the encephalitis is developed chiefly in the basilar structures.

The cause of the disease will vary also according to the intensity of the encephalitis. In some of the cases the coma deepens, the patient may linger on for two or three weeks, but finally succumbs to the disease, the entire course reminding one very much of the pyæmic forms of ncephalitis. In other patients, after the disease has lasted for a week or two, prolonged remissions and complete recoveries may set in.

As an instance of encephalitis following influenza, I wish to refer to the following case, although the possibility of a meningitis being associated with the encephalitis cannot be denied. The patient and several other members of the family had passed through the usual attacks of influenza four days previous to my first visit. The girl, eighteen years of age, had been badly frightened by the falling of a picture. That same evening she began to vomit and complained of severe headaches. The family physician, who ex-

* On the relation of the acute infantile cerebral palsies acute encephalitis, cf. Chapter XXV.

amined her, found a temperature of 103° F., and noticed at once a slight retraction of the neck. For the next few days the temperature varied between 101° and 103° F. The rigidity of the neck became more pronounced, but the pulse and respiration were not sensibly affected at any time, the former being at the rate of 90 and the respirations varying between 16 and 20 a minute. On the fourth day of the disease there was a very marked rigidity of the neck and great painfulness over the cervical and dorsal spine; excessive muscular tenderness in the lower extremities, such as is frequently found in cases of simple influenza. The deep reflexes were lively, but not excessively exaggerated. The patient was drowsy; if left to herself would sleep, but when aroused answered all questions intelligently. She presented in addition a marked paralysis of the rectus externus in both eyes. There was loss of light reflexes and sluggishness of contraction during accommodation in both eyes, but there were no other third-nerve symptoms. The papillæ were somewhat swollen. These symptoms continued, with slight changes, for two weeks; then she began to improve slowly; four weeks after the onset of the first symptoms the patient was entirely well.

These cases surely bear a strong resemblance to the ordinary forms of meningitis, and were it not for the evidence furnished by Fürbringer, Leyden, Eisenlohr, and Koenigsdorf that encephalitis is the actual condition, the diagnosis of a meningitis could be defended quite as readily.

MORBID ANATOMY.—As far as can be determined at the present time, the morbid process is a simple hemorrhagic encephalitis. The inflammatory areas are small and strictly circumscribed, and may be developed in symmetrical parts of the brain. On superficial examination the brain tissue appears hyperæmic and dotted with small red points; the areas of inflammation are a little softer to the touch than normal tissue. On microscopical examination the smaller vessels are found dilated to their utmost capacity, filled with blood, some of which has evidently been exuded into the neighboring tissue. Leucocytes, granular cells, and a proliferation of the cells of the neuroglia complete the microscopical picture. There may be slight destruction of the nervous elements, but this will depend upon the intensity of the process. The entire encephalitis is, no doubt, of microbic origin. Southard and Keene have very recently shown that it may be due to the staphylococcus pyogenes aureus.

The prognosis will naturally vary according to the intensity of the symptoms. A fatal issue is apt to follow in those cases in which the symptoms, coming on with great violence, point to an intensely septic process. If the condition of coma is reached by slow stages and remissions are observed within the first week or ten days, a favorable turn may be expected.

The treatment will consist simply in absolute quiet, the application of cold to the head and nape of the neck, and in the administration of purgatives (calomel is by far the best).

POLIO-ENCEPHALITIS SUPERIOR.—A disease due to the inflammation of the gray matter of the floor of the fourth ventricle and of the aqueduct of Sylvius, had been described by Wernicke, Thomsen, and others. This disease has been observed in the adult, and is remarkably frequent in persons

presenting the symptoms of chronic alcoholism. This special form would naturally be a rarity in children; the symptoms are, however, not unlike that form of encephalitis which has been described in connection with influenza. It is of particular interest, also, because a similar condition is at times associated with polio-myelitis. We have often referred to the close relation between the gray matter in the spinal cord and the gray matter which harbors the nuclei of the cranial nerves. We can readily understand why the same morbid process should at times affect the cerebral, and at other times the spinal portion of this central gray matter. According to the distribution of

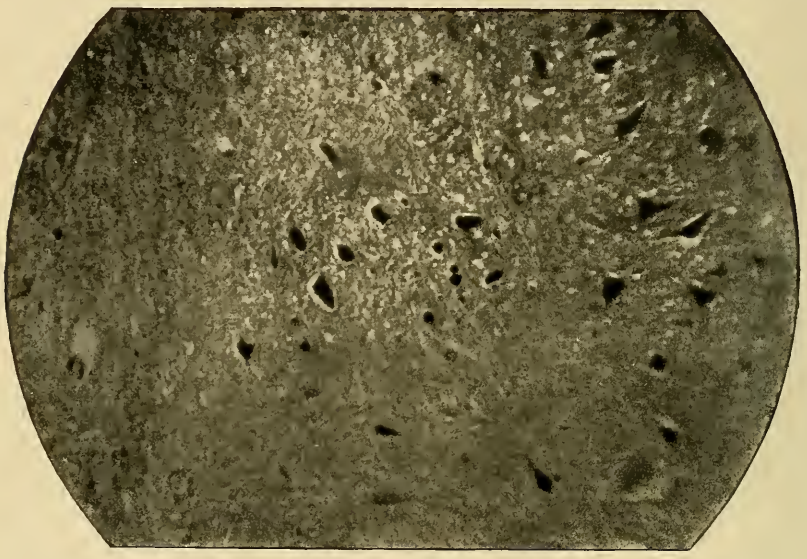


FIG. 103.—Case of Unilateral Nuclear Palsy. (Wiener.) Hypoglossal nucleus, left side. Photomicrograph from a section stained after Pal, showing some degenerated ganglion cells.

the inflammation in the region of the ocular nerve nuclei, or in the vicinity of the nuclei of the tenth and twelfth nerves, we distinguish between polio-encephalitis superior and inferior. The disease in which ocular nerve symptoms (ophthalmoplegia, partial or complete) have been associated with polio-myelitis, is termed polio-encephalomyelitis. This form has been described by Rosenthal, Seeligmüller, Guinon, myself, and others. But these diseases are, on the whole, extremely rare, and do not, as a rule, begin until childhood is past. The cases also take a more or less subacute course.

BULBAR PALSIES.*—Diseases of the pons and medulla are very rare in children; but the above discussion of nuclear diseases leads to the mention

* The author had some hesitation in discussing these diseases in this chapter; but they are too rare to warrant discussion in a separate chapter, and it did not seem desirable to add them to the sections on congenital nuclear palsies.

of progressive bulbar palsy. This disease in the adult bears a close clinical and anatomical relation to progressive amyotrophy and to amyotrophic lateral sclerosis. With the chronic diseases of adults we are not concerned, but a short allusion may be made to progressive bulbar palsy of earlier years, which is a rare disease, but, like the adult form, is due to a primary degeneration of the nuclei of the lower cranial nerves.* A case of this description, from my clinic, was carefully studied and described by Dr. Wiener.

The symptoms of unilateral bulbar palsy were discovered quite accidentally in a young man who had been under treatment by his physician for tubercular glands and pharyngitis. He presented: 1. A very marked deviation of the tongue to the right when protruded. 2. Distinct atrophy of the median portion of the right half of the tongue. 3. The faradaic response of

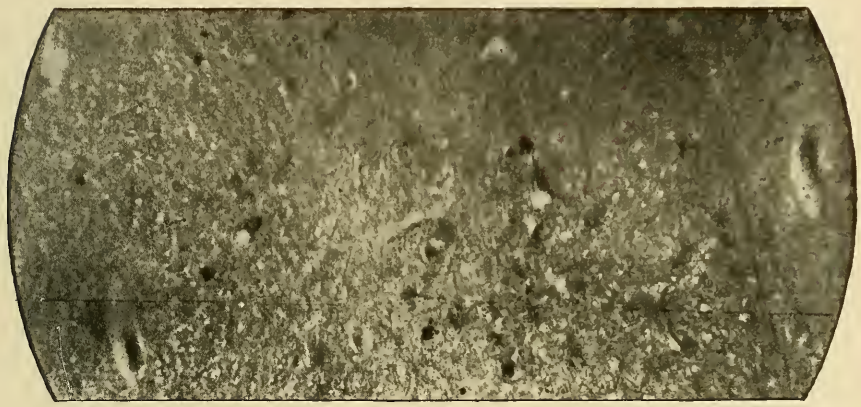


FIG. 104.—Same Case as Fig. 103. Hypoglossal nucleus, right side, showing absence and extreme degeneration of ganglion cells.

the right half of tongue was much diminished; the contractions were sluggish, there was increased galvanic excitability of the right side (K. C. C. < A. C. C.); the contractions were slow and wave-like. 4. Taste and tactile sensations were normal. 5. Deviation of the soft palate and of the uvula to the left side. On phonation the paralysis of the right side became more marked. 6. Great difficulty in deglutition. 7. Dysphonia due to disease of the right recurrent laryngeal nerve.

Other cranial nerve functions were normal. All reflexes normal.

A gradual progression of all the symptoms led to a fatal issue from respiratory failure. Examination of the brain revealed a distinct degeneration of the hypoglossal and vago-accessorius nuclei of the right side, with degeneration of the respiratory column. In the hypoglossal nucleus, the ganglion cells were greatly changed (atrophied, shrunken, and granular). The ground

* No special reference is here made to the acute and the pseudo-bulbar palsies, as both these conditions are very rare in children An acute polioencephalitis inferior in young subjects, giving rise to bulbar symptoms, has been described.

substance did not stain as readily in the right as in the left, and was less compact; the left hypoglossal nucleus was also slightly diseased (lower and outer portions). (See Figs. 103, 104.)

The symptoms of bulbar palsy (paralysis and atrophy of the muscles of the palate, the tongue, the pharynx, and the larynx) were distinctly present. In some subjects there is additional paralysis of the lips and of the muscles of mastication, and also of the upper divisions of the facial distribution. The difficulties of deglutition and the dysarthria were characteristic; all these symptoms justify the clinical designation of glosso-labio-pharyngeal paralysis. The unilateral character of the symptoms is worthy of note, but does not remove the case here recorded from the category of subacute or chronic bulbar palsies.

The disease which involves the nuclei of the motor cranial nerves is occasionally associated with spastic symptoms, with increase of the deep reflexes, and with atrophy in the upper extremities. The clinical symptoms thus bear the closest resemblance to those ofamyotrophic lateral scleros is and the latter disease may well represent an extension of the bulbar process into the spinal cord; the spinal symptoms of amyotrophic lateral sclerosis may precede the bulbar symptoms for a long period of time. All these forms represent a disease of both divisions of the motor tract.

C. H. Brown has described an interesting case of amyotrophic lateral sclerosis with bulbar symptoms which exhibited the first signs at the age of twelve years or earlier. All the facial muscles were involved; the boy could not laugh or swallow easily; speech was nasal; there was atrophy of the sterno-cleido-mastoid of the left side, and to a lesser extent of the right. Other neck muscles weak. Considerable atrophy around the shoulder girdle, of all of the arm muscles, and of the interossei and the thenar group, fibrillary twitchings and increased reflexes completed the series of symptoms. Progressive bulbar paralysis in children has also been described by Hoffman, Remak, and Londe. Eisenlohr has described a bulbar form of infantile spinal paralysis.

Charcot, Londe, Brissaud, Marie, Hoffman have described an infantile, hereditary or family form of progressive bulbar paralysis. The characteristic signs, in addition to the bulbar symptoms, were the presence of stigmata of degeneration, the involvement of the upper facial branches, the occurrence of ptosis and of a partial or complete ophthalmoplegia.

Oppenheim, Peritz, Bouchaud, Ganghofner have described an *infantile form of pseudobulbar paralysis.* This complex of symptoms is developed in association with the infantile cerebral palsies. Glosso-labio laryngeal symptoms may be combined with spastic-athetoid and diplegic symptoms. Oppenheim and Bouchaud have shown this form to be due to defective development (porencephaly) of the lower portion of the central convolutions.

CHAPTER XXIV.

HYDROCEPHALUS.

By hydrocephalus we mean an excessive accumulation of serous fluid within the cranium, either in the subdural spaces or in the ventricles. The former is termed external hydrocephalus; the latter, internal hydrocephalus. It is convenient also to divide the cases into acute and chronic forms, and to subdivide the latter into congenital and acquired hydrocephalus. As the accumulation of fluid often follows upon other diseases, we distinguish a form of secondary hydrocephalus; the term primary hydrocephalus remains restricted to the few cases in which the accumulation of fluid appears to be the sole morbid condition. Chronic hydrocephalus is by far the more important, for what might be said of truly acute hydrocephalus has already been said in connection with the primary forms of meningitis; but a few remarks will be in order, at this juncture, in relation to the subject of acute hydrocephalus, neglecting for a time the hydrocephalic condition which accompanies every form of meningitis, and particularly the tubercular variety.

Acute Hydrocephalus.—Quincke, in an able article on meningitis serosa states, that the term acute hydrocephalus should be made to cover the condition in which there is a purely idiopathic serous meningeal inflammation. In the cases which he describes, and of the existence of which there is no doubt, the symptoms point to a very constant increase of intracranial fluid in the subpial spaces and within the ventricles. Acute hydrocephalus is characterized generally by very sudden onset; occasionally the symptoms come on in insidious fashion. Fever may be entirely wanting, or, if present, rarely exceeds 103° F.; headache, rigidity of the

neck, nausea, vomiting, stupor, coma, delirium, are the symptoms present in this disease, and they are the same which we have insisted upon so frequently in connection with the various forms of meningitis. The pupils react sluggishly, and are often unequal; optic neuritis is present; convulsions may occur, but are rare, and palsies of a cerebral order may be present, but are rarely either severe or lasting. Paralysis of the external rectus muscle is as frequent as any, and this can readily be explained by increased intracranial pressure. In milder cases the symptoms soon recede, and absolute recovery may take place within a few weeks. In the more serious forms the symptoms may closely resemble those of brain tumor. The disturbance of function due to increased intracranial pressure may bring about a fatal issue. Some of these types of meningitis serosa are no doubt idiopathic, others may represent an acute exacerbation of a chronic hydrocephalus. These diseases occur rarely before the first year of life, but are most frequent between the age of one and five years. They may occur up to the age of thirty years, and even later.

Meningitis serosa may be due either to traumatism or to acute febrile disease, such as typhoid and pneumonia; in the adult the excessive use of alcohol, and the pregnant state may be responsible for the development of the disease. It is probable that this serous form of meningitis is not of microbic origin. A distinction should be made in fact between these cases and those in which some micrococci are known to be the definite cause.

Pathology.—Acute hydrocephalus may be due to a number of different causes. First, to venous stasis, such as occurs in connection with severe heart disease. Secondly, venous stasis in the larger veins at the base, due to neoplasm in the posterior cranial fossa, may also be the direct cause of acute dropsy of the brain. Thirdly, stasis in the lymphatic vessels, most frequently due to tumors in the posterior fossa, causing compression of the subarachnoid lymph spaces and of the aqueduct of Sylvius, are a sufficient cause of acute hydrocephalus. Fourth, any acute inflammatory process of the brain or of the meninges may lead to this form of meningitis. The exact symptomatology of an affection of

this nature occurring in a young person may be illustrated by the following history taken from Quincke:

A boy, nine years old, who was said always to have had a large cranium, was struck upon the back of the head. He was unconscious for a few minutes, and after the accident complained of pains in the occiput, which were generally slight, but occasionally much increased. With increasing headaches the boy grew pale, vomited, showed slight rigidity of the neck, and retraction of the head. The horizontal circumference of the head six months after the accident was 54½ ctm. No pain on percussion of the head, but pressure upon the upper cervical spinous processes was painful. Vision much diminished. The right eye had fair perception of light; with the left the boy was able to count fingers. The pupils moderately dilated, reacting sluggishly; both papillæ were atrophic. All movements were perfect. Serous fluid was withdrawn in considerable quantity on two different occasions by Quincke through a puncture in the lumbar region. Upon this there was improvement in the headaches, but sight remained permanently impaired.

CHRONIC HYDROCEPHALUS.—From this category of cases we may at once exclude those in which the increase of intracranial fluid is of a simple compensatory nature. Such an increase is found in many instances of congenital imperfect development of the brain. A skull of average dimensions sometimes harbors a very small brain, the cranial cavity being filled by an increased amount of fluid. The fluid causes a bulging of all the cranial bones. The accumulation of fluid during the intra-uterine period may increase the size of the head to such an extent as to make it a serious obstacle to normal delivery.

Deficient brain development is at times limited to one-half of the brain. An entire hemisphere may be transformed into a large serous sac, while the other half presents tolerably normal appearance. An entire hemisphere may consist of nothing more than a superficial layer of gray matter bordering upon a huge cyst which communicates with the ventricles. I have had opportunity to be present at an operation upon such a brain in which a cyst was diagnosticated, but the size of the cyst not suspected. On opening this cyst the surgeon's probe could be passed many inches forward and backward, showing that the greater part of the hemisphere consisted of this cystic mass. The boy died about two weeks after the operation, when it was

found that the entire hemisphere consisted of nothing else than a small layer of gray matter bordering a huge cystic cavity. But all these forms, however interesting they may be, have comparatively little value, and are far less frequent than those in which there is a condition of congenital hydrocephalus.

CONGENITAL HYDROCEPHALUS.—For reasons which are not well known an excessive accumulation of fluid may occur within the ventricular cavities during the intra-uterine period. The condition is found in children of absolutely healthy parents after normal pregnancies, and in families in which all other children have been entirely healthy. In some families, however, several children are born afflicted with this same trouble; one or the other may survive while the majority are either still-born or die very soon after birth. Syphilis of the parents, alcoholism, tuberculosis in the father or mother, emotional excitement of the mother during pregnancy, injuries to the mother during this same state, all these factors have been held responsible for the condition, and no doubt are operative in some of these patients, while in the majority the actual cause remains unknown. If hydrocephalus is present at the time of birth the amount of fluid present may go on increasing within the first few days, or as long as the child lives.

In this connection the observations made upon an infant whom I had occasion to examine in 1893 may be of some interest. When I saw it for the first time it was four weeks old. The mother had given birth to one other girl, three years previously, that had remained healthy and showed no signs of internal hydrocephalus. The father was a man of remarkably vigorous health. During this second pregnancy the mother menstruated regularly. On account of the size of the head labor was extremely difficult, but the child was uninjured when born. The measurements which were taken at three different occasions will show the rapid increase of the fluid, as indicated by the rapid increase in the head measurements:

	Aug. 6. Inches.	Aug. 8. Inches.	Aug. 10. Inches.
Horizontal circumference.....	20⅜	21½	22
Naso-occipital measurement.....	13½	12½	14½
Bin-auricular measurement.....	12½	13½	13

On the 9th, between the second and third measurements, a large amount of fluid was withdrawn by tapping the fontanelles, but it will be noticed that

the fluid was rapidly replaced. The child lingered on for another week and then died, after having become greatly emaciated. More than a week before its death it could not be fed by the mouth or per rectum.

But all cases of congenital hydrocephalus do not take such a serious turn, and in some of them the accumulation of fluid ceases and a tolerably fair and even normal mental development may ensue. The protruding occipital bones clearly visible on so many bald heads point to a moderate amount of internal hydrocephalus in the earlier years of life. Some of these may have been acquired in the earlier period of life and are not necessarily congenital in origin. The fluid accumulates most readily within the lateral ventricles, both in the anterior and in the posterior horns. The aqueduct of Sylvius may be distended into a funnel shape by the increased fluid, but the fourth ventricle, as a rule, suffers very little. The brain is compressed by the accumulation of fluid, and is often so flattened and thinned that its thickness is not greater than that of ordinary thick paper. The white matter seems to yield more readily than the gray, and I have been struck, in a number of different brains, by the fact that the function of the parts remained normal in spite of the extreme thinness of the cerebral tissue. Thus in one instance in which both occipital lobes were reduced to the thickness of paper, sight had remained entirely normal up to the age of two years, at which time the patient died. The membranes are, as a rule, normal, or else show only very slight thinning, and on microscopical examination only the slightest traces of acute inflammation or none at all. The choroid plexuses are generally thickened, as is also the limiting membrane of the ventricles.

The fontanelles bulge and pulsate distinctly. The configuration of the skull is modified by the excess of internal hydrocephalic fluid. If the hydrocephalus is congenital, or if it has occurred in very early life, the bones yield to the increased pressure within and the skull becomes distended in all directions. In keeping with the greater accumulation of fluid in the anterior and posterior horns of the ventricles and the protrusion of corresponding parts of the brain, the greatest amount of bulging is observed in the frontal and occipital bones respectively. The effect of the excessive ac-

cumulation of fluid in the posterior horns of the ventricles results in a great bulging of the occipital bones, with marked increase in the transverse occipital diameter of the skull. If the child survives, the congenital internal hydrocephalus gives rise to a number of symptoms pointing to deficient cerebral development. On account of the size and weight of the head the child is rarely able to carry the head unsupported. As it grows older it may be able to sit in a chair, but the head generally inclines to one side or the other, or forward, in which case it may rest upon the chest. Defective mental development is present in the majority of these cases, ranging between total idiocy and varying degrees of imbecility; some of the children acquiring a slight use of language and others learning to utter only a few words. Wyss states that of forty-five hydrocephalic children only five were able to attend school. Cerebral palsies with typical spastic contractures are quite frequent, and convulsive seizures amounting to a chronic epilepsy often add to the burdens of the child, and to the distress of the mother who is to care for it. In some cases there is total blindness, due to optic-nerve compression, but the most surprising feature of all is the remarkable preservation of the brain functions.

Acquired Internal Hydrocephalus.—Chronic internal hydrocephalus, not of congenital origin, is a rare disease, and it is at least in rare instances only that the diagnosis of such a form of internal hydrocephalus can be made, and if made, the cause which gives rise to it is so far more important that the internal hydrocephalus is a mere incident and does not deserve much notice. This acquired internal hydrocephalus may be of primary origin or it may be due to some mechanical obstruction causing venous stasis. The primary form may be a symptom of severe general anæmia, and under such conditions it is a counterpart of serous transudation occurring in various cavities of the body. It may also occur in children who have rickets, or in those whose skull is sufficiently yielding to permit enlargement of the cranial cavity. The increase of fluid is to a certain extent compensatory in such cases.

Internal hydrocephalus due to mechanical obstruction of the veins of Galen, was first described by Whytt. The commonest form of obstruction is that due to tumor in the posterior fossa. Through such obstructions the foramen of Majendie may become occluded and dilatation of the third ventricle will result; in the case of occlusion of the foramen of Monro, a distention will take place in the lateral ventricle, but increased secretion of serous fluid may be sufficient to cause distention of all the ventricles without any

obstruction whatever. On the other hand, a meningitis may be sufficient to occlude these small openings connecting the ventricles with one another, and may thus become the actual cause of an internal hydrocephalus. This special cause of internal hydrocephalus is of interest only in those comparatively few cases which survive a severe attack of meningitis. A slight increase of intracranial fluid is present in almost every case of intracranial growth, and is responsible for many of the general cerebral symptoms accompanying such diseases; but as we said above, the recognition of tumor is so much more important than the diagnosis of an accompanying hydrocephalus that the latter loses very much in importance.

The symptomatology of the acquired form of internal hydrocephalus differs in no respect from that of other chronic forms except that the symptoms develop with some suddenness; vertigo, dizziness, stupor, coma, convulsions, contractures, and amblyopia, preceded possibly by a condition of hemianopsia, are the symptoms which should be looked for in these patients. It stands to reason that if the internal hydrocephalus originates at the time when the bones are still yielding, the injury to the brain and the symptoms resulting therefrom will be far less marked than they will be if it occurs after complete ossification of all the sutures and bones. After the sutures have been closed the increasing hydrocephalus may force them open again. I have not observed this in any of the primary forms of hydrocephalus, but have seen it occur in a child of four years, who died of cerebellar tumor with enormous internal and external hydrocephalus. The acquired internal hydrocephalus runs very much the same course as the chronic form, and is very apt to terminate fatally in a few weeks or months. Moderate cases may survive for a longer period of time.

The diagnosis of all forms of internal hydrocephalus depends entirely upon the association of an enlargement of the cranial cavity with symptoms which point to an increase of intracranial pressure. The exact form of hydrocephalus, whether it be congenital or acquired, can be diagnosticated only with reference to the history of the individual. In endeavoring to determine whether the hydrocephalus is internal or external, we should remember that the external hydrocephalus is more apt to be congenital and to be associated with symptoms of defective mental development; and furthermore, that bulging of the frontal or occipital bones only accompanies internal hydrocephalus; whereas, a general enlargement of the skull in every diameter, with bulging fontanelles and sutures forced apart, indicates the presence of external hydrocephalus, which is in most cases associated with distention of the ventricles as well. But in many children the distinction between external and internal hydrocephalus is difficult to make, for the internal hydro-

cephalus may have so thinned out the cerebral substance as to bring the ventricular fluid very near the surface.

Further difficulties of differential diagnosis which arise occasionally are to distinguish from hydrocephalus the enlargement of the skull due to rickets, to syphilis, or to thinning of the skull resulting from other causes. In the case of rickets, the entire history of the disease will give a clew, and other symptoms of rickets will be present and will indicate the true nature of the enlargement of the head. But both rickets and hydrocephalus are not infrequently associated with each other. An increase in the size of the skull may simulate the condition of hydrocephalus. Such thickening is extremely rare, but is met with in the subjects of hereditary syphilis. I have seen this but once, in a boy, aged six years, in whom there were other signs of hereditary syphilis; yet the skull is never so much enlarged as in the extreme cases of hydrocephalus, and generally impresses one as being unusually hard.

PROGNOSIS.—The prognosis of hydrocephalus is always serious, but depends greatly upon the original cause. Fortunately a goodly number of children with varying degrees of hydrocephalus not only live a number of years, but get well. The congenital cases are more rapidly fatal than the acquired form, and the prospects of a normal mental development are always slight, though remarkable exceptions may occur. Measurements of the skull taken at regular intervals will enable the physician to gauge the rate of increase in the accumulation of fluid, and will help him to prognosticate the future course of the disease.

TREATMENT.—In the milder forms of hydrocephalus, the iodides and the preparations of iron will do good service. In the severer forms of hydrocephalus the attempt should be made to absorb some of the fluid which is present in excess by the use of the mercurials and diuretic measures. I cannot state that I have seen any favorable results following upon this course of treatment, though some temporary relief has been afforded in a few cases. The treatment is, however, a thoroughly rational one. In almost every form of acute and increasing hydrocephalus, however little faith he may have in the efficacy of the method, the physician or

surgeon will be tempted to follow the example of Keen, Kocher, Bergmann, Chipault, to tap the ventricles, and thus drain away some of the fluid. The fluid is replenished so rapidly that very little good can be effected in this way, and if it is drained off suddenly the danger of collapse is very great. Henschen's statistics have been most discouraging. At the present day, lumbar puncture will take the place of this procedure, but is, of course, useless in those cases in which the communication between the ventricles and the cerebro-spinal canal is interrupted. Instead of draining away a large quantity at one time, it seems much more rational to attempt some method of gradual drainage.

Every other treatment, including the injections of iodine, the application of elastic bands about the head, and of compression of various kinds, is useless. I have often attempted to reduce the skull by applying bandages more or less firmly around the head, but the result has in every instance been equally unsatisfactory. If the skull is firmly compressed the internal pressure is naturally increased, and if the compression is not firm enough the bandages will do no good.

CHAPTER XXV.

INFANTILE CEREBRAL PALSIES.

(SPASTIC HEMIPLEGIA, DIPLEGIA, PARAPLEGIA.)

THE investigators of recent years have with few exceptions endorsed the classification of the cerebral palsies, as given in the first edition of this book. We may divide these palsies into three groups according to time of onset—during the intra-uterine period, during labor, or after birth. A clearer understanding of all these disorders will be reached by discussing them together, comparing and contrasting the chief features of the one series with those of the other groups.

The subject is not a new one, yet the various forms of cerebral palsies did not attract the attention which they deserved until two decades ago. A clear account of their clinical symptoms was given by Von Heine in the second edition of his book on "Infantile Spinal Palsies" (1860). Heine distinguished between the typical cases of infantile spinal paralysis and those of a purely cerebral character. Before his day some of the older French authors —Cazauvielh, Breshet, Cruveilhier—had studied the atrophic changes in the brains of children who had been afflicted with various forms of palsy. These writers were interested in the anomalies of brain structure rather than in the clinical features presented by such cases. In 1842 Henoch published a dissertation entitled "De Atrophia Cerebri," in which he described the brain condition associated with infantile cerebral hemiplegia. The chief advances were due, however, to Cotard, a pupil of Charcot, who in 1868 published "A Study on the Partial Atrophy of the Brain," in which he analyzed the different morbid processess to which such atrophy might be due. Cotard was first also to recognize the importance of traumatic encephalitis as a factor in the causation of these conditions. A special study of brain defects was made by Heschl, who introduced the term "porencephaly," and by Kundrat, who in 1882 published a monograph on this same subject, in which he distinguished between congenital and acquired porencephaly, and attempted to explain all these conditions by attributing them to a form of anæmic necrosis. In 1883 Audry published a collection of one hundred and three cases of this

peculiar condition. Lobar sclerosis, a condition to which Cotard had called attention, was made the special subject of papers written by Bourneville, Richardière, Jendrassik, and Marie. The last author introduced a decided advance in the discussion of the subject by connecting this lobar sclerosis with changes in the blood-vessels, and still the character of the initial affection remained rather doubtful. Quite apart from all these French and German authors, Little, in 1853, had studied carefully the spastic palsies and rigidities of children, and showed that they were largely due to protracted labor and premature delivery. The conditions due to these two causes have since been known, particularly among German and French authors, as "Little's Disease." (Cf. page .)

A fresh impetus was given to the study of all these palsies by the lecture of Strümpell, delivered in 1884, who declared that acute infantile spastic hemiplegia was due to a primary acute encephalitis affecting the gray matter of the cord. He insisted upon an analogy between this acute cerebral and the acute spinal palsies, and in order to fortify the resemblance between the two sets of disease, proposed the term "polio encephalitis" as the cerebral counterpart of "polio-myelitis." The theory advanced by Strümpell was ingenious; there is more clinical than anatomical evidence to support it. We can not, even at this day, agree with Leube, who states that "hemiplegia in young children should in the first instance be considered to be the result of a former acute encephalitis." The truth is that encephalitis is the rarest cause of these palsies, and should not be considered at all, unless every other morbid state can be safely excluded. But of this later on.

Strümpell's lecture was the starting-point of the innumerable studies which have led to the clearer recognition of the true pathology of these disorders.*

The brain palsies of children are far more frequent than they are generally supposed to be. In the Hospital for Ruptured and Crippled in this city, Dr. Townsend, at my suggestion, was kind enough to tabulate the cases of spinal and cerebral infantile palsies which presented themselves for treatment at that institution for a definite period of time. His statistics show that during that period ninety-one patients with infantile cerebral palsies, and one hundred and forty-two cases of infantile spinal palsies were received for treatment. I had, during a period of five years, seen over three hundred cerebral spastic palsies, but this may be due to the fact that many physicians knowing my interest

* Among the many writings the most prominent are those of Bernhardt, Wallenberg, Kast, Moebius, Feer, Freud and Rie, and Oppenheim, in Germany; of Marie, Cotard, Audry, Gibbotteau, in France; of Ross, Hadden, Gowers, and Ashby, in England; in this country numerous articles have been contributed to this subject by Weir Mitchell, Sarah McNutt, Lovett, Osler, Peterson, Sachs, Starr, Spiller, and Dercum.

in the subject were at special pains to refer such cases to me.

Whatever the time or manner of origin may have been, all cases of cerebral palsy in children are so similar in many respects that we may attempt to study the symptoms common to all. These various forms of cerebral palsies are observed most frequently in the earlier years of life, from the time of birth up to the age of ten years, and even later, but by far the greater majority of them occur during the first three years of life. The paralysis is of the distinctly spastic order and according to the distribution of the palsy we may distinguish between a hemiplegia and diplegia (double hemiplegia) or paraplegia. Monoplegias are relatively rare. The rigidity of the muscles, the contractures resulting from the same causes, and increase of all the deep reflexes are the constant accompaniment of these palsies. Coma and convulsions occur in the initial stage of the acute forms, and the convulsions at least are often repeated during the later stages of the disease. The cerebral palsies of children are more commonly associated with coma and convulsions than are those of the adult; the former are generally due to cortical processes, the latter to intra-cerebral conditions. The frequent repetition of convulsions is equivalent to the establishment of an epilepsy which may continue throughout life, and the same changes which have caused the epilepsy may also be responsible for the defective mental development, which may range from weak-mindedness to marked imbecility and complete idiocy. Disturbances of motion, associated, ataxic

FIG. 105. — Left Hemiplegia. Onset in Second Year. Contractures of arm and fingers. (See also Fig. 75.)

athetoid, choreiform, and even cataleptic movements, occur quite often in connection with these diseases. All of them may be grouped under the general heading of post-paralytic (not necessarily post-hemiplegic) disturbances. Aphasia, so common in apoplectic disorders of the adult, is a rarer complication in infantile palsies.

Among the negative symptoms which are of the greatest importance in attempting a differential diagnosis between the cerebral spastic and the spinal palsies, we may mention the entire absence of changes in the electrical reactions and the development of only slight atrophy in association with the palsy. Disturbances of sensation are rare, but as these are absent in the spinal forms as well, they help little in attempting a differential diagnosis between these two important series of diseases.

Let us now take up the symptoms *seriatim*, and in doing so we shall be able to develop the many interesting features of infantile cerebral palsies.

Distribution of Paralysis.—The following analysis of 225 cases is based upon the collections made by Peterson and myself :

	Males.	Females.	Total.
Right hemiplegia	51	30	81
Left hemiplegia	40	35	75
Diplegia	21	18	39
Paraplegia	22	8	30
Total	134	91	225

Monoplegias are so rare that they scarcely enter into the consideration of the problem. From the above statistics it will be inferred that right and left hemiplegias are more frequent that the bilateral forms of palsy, which is equivalent to saying that unilateral cerebral processes are more common than double cerebral lesions. There is no difference more striking between the adult and the infantile palsies than that implied in the relatively large percentage of cases of cerebral diplegia and paraplegia. In the adult a bilateral cerebral lesion is a great rarity, but in children diplegias constitute about fifteen per cent. of all the cases, and paraplegia about fourteen per cent. This difference between the adult and infantile brain is due first of all to the fact that both hemispheres in children are exposed to the same external injuries at birth, and even if the disease is acquired later in life the two hemispheres seem often to be affected simultaneously, and much more frequently than in later years.

In my own statistics boys were affected a little more often than girls, but the difference is hardly great enough to be made much of, and Gowers has found just the reverse to be true. Osler found that girls and boys were affected in about equal proportion.

The onset of a cerebral palsy will vary naturally according to the three groups which we have established. It would seem entirely superfluous to make any special remarks regarding the time at which the prenatal palsies begin. It might be sufficient to say that they begin during the intra-uterine period, and that such children are born palsied. It is a matter of fact that the symptoms of such intra-uterine cerebral defects are not always manifest at birth, and indeed a number of months may pass before it becomes evident to the physician that the child's cerebral condition is not a normal one. A very fair percentage of cases which appear to begin during the first years of life could properly enough be classified among the prenatal palsies, and if in any child the first symptoms of a cerebral palsy are noticed several months after birth, and yet the period of labor was entirely normal, I should be inclined to classify that case rather among the prenatal palsies than among the birth palsies.

FIG. 106. — Right Hemiplegia, with Contractures and Retarded Growth of Arm. Onset of disease at eight years of age, following typho-malarial fever.

The birth palsies begin naturally enough with the period of labor. The history in these cases clearly shows that the labor was either excessively prolonged, or that an instrumental delivery was resorted to, in which case the brain has evidently suffered mechanical injury. Premature delivery is responsible for many cerebral palsies; but the symptoms may not be fully developed until months after birth.

The age of onset in the acute or acquired forms varies considerably, and yet the chief statistics agree in this that fully two-thirds of all the cases begin in the first three years of life. Some maintain that by far the largest proportion begin during the first year, but I am not inclined to accept this view, since such a list would include a large number of cases which are more properly congenital, although the symptoms do not become manifest until some time after birth. Among a total of one hundred and forty cases, Peterson and I found that but five began in the eighth year, and four cases

between the ages of fourteen and fifteen years, but these were cases of hemiplegia, and it is of some interest to note that no patients with diplegia or paraplegia were observed after the third year, showing that the causes which lead to double cerebral lesions, and possibly the condition which permits double lesions, cease with the very earliest years of life. The majority of diplegias and paraplegias are either congenital or occur during the first year, which can be easily explained by the fact that the injuries sustained during labor are largely responsible for these special types.

ETIOLOGY.—The true etiology of the prenatal cerebral palsies is often rather mysterious. We have to resort to vague statements that they are due to hereditary taints, and as a matter of fact such palsies occur frequently enough in families with a decided history of neurotic taints of one kind or another, with a history of hereditary epilepsy, or of hereditary insanity. I have a number of times observed such children, in whom an hereditary taint was present in the families of both father and mother. The next most important factor is unquestionably the occurrence of some accident or injury to the mother during the period of pregnancy. This was to be traced distinctly in a number of cases of my own. The manner in which such traumatism may do actual injury to the brain of the child, was clearly shown by a case which was cited by Cotard. The mother of the child sustained a blow against the abdomen during pregnancy. The child, which was still-born three months after the injury, exhibited extreme contractures of both left upper and lower extremities. On examination of the brain an old lesion was discovered in the right hemisphere in the vicinity of the lateral ventricle.

Illness of the mother during pregnancy, exhausting fevers, particularly such as accompany pneumonia and typhoid, uræmic convulsions, and severe fright, are other causes which have been made out in cases of prenatal palsies.*

* Osler reports (in Teratologia, vol. ii., No. 1), upon the case of a young woman who died of typhoid fever. "The uterus contained a fœtus, apparently about six months old. . . . The brain was very soft; the right hemisphere was normal, but in the left . . . was a cavity with ragged, irregular walls, containing a large, recent clot, which had broken through the ganglia into the lateral ventricle of the same side. . . . No special changes were noted in the arteries."

Hereditary syphilis does not play an important part in the etiology of prenatal palsies. Erlenmeyer, Osler, Oppenheim, Koenig, and others mention it. As a matter of fact I have been able to convince myself of the influence of syphilis in but a single one of the many cases which I have seen. The fact that syphilis of the parents so frequently leads to still-births may account in a measure for the small part played by this special affection in these diseases.

The etiology of birth palsies is a very simple one. It is surprising to note how much pressure the brain and skull will tolerate without injury, but it is natural that harm should occasionally be done. Asphyxia at birth has since Little's day been considered a most potent factor. My own studies in this matter, which were based upon a very careful collection of statistics, have proved that tedious labor is a more frequent and a more disastrous factor than instrumental delivery. Moreover, these birth palsies occur most frequently in first-born children.

The prolonged compression of the skull during the last months of pregnancy would seem to have exercised considerable influence in this direction, but, of course, the chief damage done is done during the period of actual labor. If physicians were more confident of the safety of the forceps and of their own skill to apply the same, protracted labor would not be as powerful an etiological factor as it is at the present time. There is, therefore, a distinct inference to be drawn from these facts, and a word of warning should be uttered to the obstetrician that, other things being equal, and, above all, the life of the mother not being in danger, it is wise to curtail the period of labor as much as possible, and not necessarily to wait until the child's heart action becomes feeble. Many children might have escaped idiocy and epilepsy if the period of labor had been properly managed.

Peterson is of the opinion that some, though a very small percentage (of birth palsies) may be due to cord lesions at birth. The same view has more recently been advocated by Dejerine. H. R. Spencer, among one hundred and thirty still-born children, found hemorrhage into the spinal canal and cord in thirty. Little suspected that "spinal meningitic and myelitic affections may play a considerable part in the phenomena of spastic rigidity"; he also refers to a case of spastic rigidity, reported by Dr. Marion Sims, in which a coagulum of blood was found occupying the whole length of the spine.

Acute cerebral palsies may be due to a variety of causes. The acute infectious diseases play a very important rôle; measles, scarlatina, typhoid, small-pox, and even a tonsillitis

have been the precursors of such palsies. The same have also occurred after pneumonia and whooping-cough, but in the case of the latter it is questionable whether the palsy is not as often due to mechanical injury during a spasm of coughing as to the effect of the toxic agent. Fright—which Freud has interpreted to be equivalent to a psychic trauma—is an occasional cause of acute cerebral paralysis, but actual traumatic violence to the skull is a much more powerful factor. These palsies also occur after simple or cerebro-spinal meningitis, after an exhausting gastro-enteritis, and after other severe fevers; but, of course, there is always the danger of making a *post hoc* a *propter hoc*. Among ninety-one cases of acquired cerebral palsy the exact cause could not be ascertained in twenty-seven.

There has been a tendency, particularly among French authors, to claim that all cases of acquired cerebral palsy were due to acute infectious diseases. This seems to me to be straining the point altogether too much, nor is there much evidence that acute cerebral palsy in itself represents an acute infectious disease, as Strümpell claimed some years ago. As a matter of fact only relatively few of the cases of acquired cerebral palsy begin with fever, coma, and convulsions, the very symptoms which Strümpell thought most characteristic of polio-encephalitis. The writer has departed from his former views to the extent of conceding that a small number of the acute cerebral palsies of children may be due to an hemorrhagic (infectious?) encephalitis.

The importance of convulsions as an etiological factor in acute cerebral palsies has given rise to some discussion. Freud and Rie are of the opinion that convulsions denote the onset of the cerebral process, but that they never hold a causal relation to the palsy which results from the cerebral lesion. I grant that in a very large number of cases this statement of the German authors is tenable, but in many others the palsy seems to be a more or less immediate result of the convulsive seizure. This view has been urged by Osler and myself. Anyone who has observed the marked disturbances of circulation at the acme of convulsions can readily conceive how easily a blood-vessel could burst during this period as a result of excessive stasis. This does occur, moreover, not only in children, but even in adults. I have the brain of a girl aged seventeen, who had had chronic epilepsy for years, but whose general health was not affected by the convulsive seizures; after the

last attack which she had she became somnolent, comatose, and died after three days. Her brain was covered by a large subpial extravasation, which almost completely covered the entire left hemisphere and part of the right. If such an occurrence can take place in the brain of a girl of seventeen there is no sufficient reason why similar accidents of lesser extent should not happen frequently in the case of children. Ashby, not long ago described the brain of a child, twelve years of age, in which a number of old cysts were found which were probably the result of hemorrhages occurring during convulsions. There is clinical evidence also which cannot be neglected which tends to show that the convulsions constitute a decided danger to the child, and mark a turning-point in its entire career. I have still under my observation a child, now eighteen years, which I knew from the time of its birth, and which was entirely normal until the age of fifteen months. It had begun to stand, to walk a little, and had acquired some speech. It was in every way a healthy and mentally vigorous child. At that age it was stricken down with chicken-pox, and had a single convulsive seizure, with a marked rise of temperature at the onset of this acute infectious trouble. From that time on the child's mental condition changed; it lost its speech, and to this day, although it has grown physically, its mental condition is that of complete idiocy. It would be a very remarkable instance indeed if an acute cerebral process had come on at exactly the same time as the chicken-pox.

FIG. 107. — Congenital Diplegia — "Frog Girl." Double talipes equino varus; athetosis of left hand; right arm also weak; intelligence good.

THE FORM OF PALSY.—Hemiplegia, diplegia, and paraplegia are the usual forms of cerebral paralysis in children. Monoplegia, which we might expect on theoretical grounds, is, as a matter of fact, extremely rare. (Figs. 105–108.)

The leg evidently recovers very much more quickly than the arm, as in adult hemiplegia, and for this reason it is a very great rarity to find a monoplegia of the leg with few symptoms in the arm of the same side. Under the heading diplegia we may classify all those cases in which both halves of the body have been involved, and it is better to attach the greatest importance to this bilateral character of the palsy, even though the palsy be incomplete, for the bilateral affection points to a double brain lesion, and that is the salient point in every such patient. In diplegia the legs may often be

much more affected than the arms, and some authors might be inclined to classify such palsies under the category of paraplegia. In a number of instances there has been a very complete spastic paraplegia associated with athetoid or choreic disturbances of motion in the upper extremities. This proves that the upper extremities were at one time involved, and for this reason I prefer to denote such conditions as an incomplete diplegia, or diplegia with partial recovery. Diplegias or paraplegias, in short double cerebral palsies, are, in the large majority of instances, due to a prenatal lesion, or to traumatism during labor, but I have been able to satisfy myself over and over again that both diplegias and paraplegias do occur in acute cerebral cases, and for this reason, in addition to others, I must protest against Strümpell's views of making infantile hemiplegia synonymous with the acute cerebral palsy of children.*

The involvement of the face is a matter of some interest. In children that are observed long after the onset of the disease the face appears to be entirely normal, but in fully twenty per cent. of the acute forms which I have had an opportunity of examining, facial palsy was present in the earlier stages of the disease. I have never observed a double facial palsy in cases of diplegia. The reason of this is not far to seek, for the peculiar position of the facial centre is such that freshly exuded blood would not be apt to adhere to this part of the brain. Since the publication of my own articles on this subject, and those of Freud and Rie, Koenig has called attention to the occurrence of mimetic facial palsies in children. The cases which he cites are very convincing, but this special form of facial palsy is surely very rare, or else it would scarcely have escaped the observation of so many authors.

Aphasia is at times associated with acute cerebral palsy. It is invariably motor, not sensory in character. Of course aphasia will be developed only in those children who have acquired articulate speech before the onset of the cerebral palsy. This excludes from the list all diplegias and paraplegias which have come on before or during birth. There may be defective development of speech, but no aphasia in any true sense of the word. I have the records of at least seventeen children with hemiplegia and undoubted aphasia. Of these seventeen, ten occurred with right hemiplegia, and seven with left hemiplegia. Eight of these seventeen were observed by me in private practice, and of these eight, five have been distinctly aphasic, and three of the five were cases of left hemiplegia. It is of interest to note the relatively large proportion of aphasia in children with left hemiplegia. As we grow older the left hemisphere evidently obtains the upper hand, and after the earlier years of life we become, for all practical purposes, more and more left-brained. Bernhardt has come to the same conclusion that aphasia in children accompanies right as well as left hemiplegia. Aphonia has been recorded by some.

Inasmuch as the cerebral palsies are due to lesions which involve other parts of the brain as well as the motor areas, it is not unreasonable to look for additional symptoms in these cases. Thus Freud was the first to call attention to the fact that hemianopsia was at times associated with the brain

* The experience of more recent years has fully corroborated these opinions.

palsies of children. Oppenheim, Peritz, and some French observers have noted the occurrence of peculiar reflex actions—"Feed reflex"; also an increased "motor acoustic reaction." The latter is identical with the "hyperacusis" to which I directed attention many years ago. (See Amaurotic Family Idiocy.)

The rigidities and contractures deserve especial notice. Some degree of contracture is present in fully seventy-five per cent. of the diplegias and paraplegias. The contractures occur early after the onset of the palsy, and evidently are developed much more easily than in the cerebral palsies of the adult. It is interesting in this connection to refer to the case of Cotard, in which the palsy was due to an intra-uterine lesion, and the child was born with contractured extremities. The contractures may vary greatly in degree and in the number of joints affected. The flexors and pronators of the arm, the flexors of the legs and of the feet, are most frequently affected. In the cases of diplegia and paraplegia there is, in addition, a contracture of the adductors of the thighs, which is responsible for the peculiar cross-legged position of the legs and for the cross-legged progression if the child is at all able to walk. All these contractures give the child a characteristic position and gait by which we can recognize the trouble at first sight. Pes equinus or pes equino varus is the most common deformity of the foot. In a few cases an equino

FIG. 108. — Case of Spastic Diplegia. Attempt to walk; cross-legged progression; rigidity and paralysis of legs and of right upper extremity; left upper extremity weak.

valgus is present. If the upper extremity is contractured the arm is, as a rule, in close juxtaposition to the trunk, flexed at the elbow, and the hand is generally in a position of extreme flexion, the fingers often being firmly pressed into the palm of the hand. (See Fig. 106.)

The gait varies much and is dependent both upon the paralysis and the degree of contracture. Many cases of hemiplegia in children have exactly the same walk as the adult hemiplegic patients have, but the peculiar cross-legged progression in cases of diplegia and paraplegia is characteristic of these infantile palsies, and has no counterpart in the cerebral palsies of the adult. In one case the contractures of the hip and knees were so extreme that the girl, who was otherwise well developed and bright, could walk only by skipping in the manner of a frog. She was baptized in my clinic as the "Frog girl." (Fig. 107.)

The reflexes are almost invariably exaggerated, as can be expected from the fact that the lesion is in the first division of the motor tract. In a few instances only have I found the knee-jerk either normal or diminished. Babinski's reflex can be elicited in most cases, and Oppenheim's tibial reflex in some. (Dorsal flexion of foot and toes following upon stroking of inner side of leg.) In one child seen at my clinic the reflexes were increased in the upper extremities, and decreased in the lower, although the case was one of right hemiplegia; on closer examination we discovered that two years after the onset of the cerebral palsy the child suffered an attack of polio-myelitis involving the right leg. This is, by the way, the only patient I have seen with a spinal lesion complicating cerebral disease. In some cases the reflexes appear to be absent, but this is generally due to a very marked contracture of the opposing muscles, which do not permit the excursion of the leg. Under such circumstances, however, even though the knee-jerk be wanting, the anterior thigh muscle can be seen to contract upon the tapping of the tendon. The ankle clonus and the triceps reflex are often inhibited for the reasons just stated. In other individuals, the reflexes are usually so much exaggerated that a single tap of the tendon is sufficient to produce clonic contractions of the muscle.

The *post-paralytic disturbances of motion* constitute a very characteristic feature of cerebral palsies in children, and they follow with much greater regularity upon the infantile cerebral lesions than they do upon the brain lesions of the adult. That they are by no means uncommon in the brain lesions of the adult, I had occasion to observe several times, notably in an old woman, with a lesion in the crus, whose paralyzed arm performed the most violent ataxic movements. In children these post-paralytic disturbances of motion occur in fully one-third of all the cases.

From a collection of statistics based upon one hundred and fifty-six cases of hemiplegia, and thirty-nine cases of diplegia, it was evident that athetoid, choreiform, and associated movements, are the most frequent disturbances to be noted. The choreiform and athetoid can generally be differentiated from one another, but in some cases there may be a combination of both. The athetoid movements are of especial interest, inasmuch as they are observed almost invariably in connection with the cerebral palsies, and it is questionable whether athetosis ever occurs as an independent disease, as was claimed by Hammond, who, however, deserves all the credit for the first vivid description of these peculiar movements.

The associated movements are often developed to a startling degree, the paralyzed hand imitating all the movements of the normal hand. Thus a patient who is asked to button or unbutton his clothes, will imitate all the movements of the sound hand, by the paralyzed hand, although the latter may be held in mid-air.

The choreiform movements occurring after cerebral palsies deserve some special notice, for such conditions are at times mistaken for cases of ordinary chorea. I have been consulted in not a few instances for what was supposed to be incurable St. Vitus's dance, which on examination has proved to be a form of infantile cerebral palsy, in which the palsy had largely disappeared, but the choreiform movements were left as evidences of the former more serious disturbance. In such children the presence of paralysis or of contractures, however slight, and the exaggeration of the reflexes in the choreic extremity, will be sufficient to indicate the nature of the choreic movements.

Atrophy of the muscles plays an entirely negative part in the vast majority of infantile spastic palsies. Quincke, Borgherini, Darkschewitsch, Eisenlohr, and others, have proved beyond a doubt that a cerebral muscular atrophy does at times occur, but it is still questionable whether this is due to a lesion of the trophic centres in the brain, or whether the spinal gray matter has not in some way been involved. I have observed a considerable atrophy of the muscles in a few cases of diplegia and paraplegia. In one patient with paraplegia the atrophy was so extreme that the case would have appeared to have been one of spinal palsy if the presence of the reflexes and the electrical reactions, as far as any could be elicited, together with the entire history of the disease, had not pointed unmistakably to a cerebral lesion. While a true muscular atrophy does not occur, as a rule, the affected limb remains stunted in growth, and after a few years is considerably shorter than its fellow of the opposite side. Ibrahim and others have reported hypertrophy of the paralyzed extremities, together with athetosis, but it has not been my good fortune to see more than one such case.

Remak has reported a case of luxation of the shoulder-joint in an infantile cerebral hemiplegia, which he attributes to unequal innervation of the muscles surrounding the shoulder-joint.

A distinct asymmetry of the body is present in many of the cerebral palsies, and to this we may add the asymmetric development of the skull, which has been shown to be present, by the careful measurements of Peterson and Fisher, in the majority of the subjects of infantile spastic hemiplegia; but the skulls of such children are often not only asymmetrical, but also deformed in various ways. Some of them are microcephalic, others leptocephalic, dolichocephalic, etc. Microcephalus is frequent enough to justify me in cautioning the surgeon who may be ready to operate upon all such skulls that a small skull may harbor a diseased brain. Ibrahim has written on the association of microcephalus with infantile spastic diplegia, and believes that this association points to the prenatal origin of these diplegias.

FIG. 109.—A Child with Congenital Diplegia and a Microcephalic Skull; Strabismus and Idiocy.

The most serious of all the symptoms associated with infantile cerebral palsy is epilepsy. A number of children have been brought to me with the diagnosis of epilepsy, pure and simple, in which this condition was the outcome of the same cerebral disturbance which gave rise to the palsy. The palsy may disappear, or may be so slight as to give little inconvenience to the child, whereas the epilepsy unfortunately remains. This association is so common that I am inclined to the view that a fair percentage of the cases of supposed genuine epilepsies may be attributed to cerebral lesions which occurred in early childhood, and have given rise to cerebral palsies. I am glad to see that this view has been quoted approvingly by a number of authors, among others by Freud, who has unquestionably given this entire subject the most careful study. A single case will bring out the truth of this very clearly.

Some years ago I was asked to see a girl, seventeen years of age. A number of able physicians had treated her for genuine epilepsy. Upon examination of the girl I was astonished to find that the left extremities were weaker than the right, and that the deep reflexes of the left upper and lower extremities were considerably exaggerated. Upon questioning the mother closely I discovered that four years previously the girl had had a slight apoplectic seizure, after which the convulsions first appeared. The palsy had diminished to such an extent that everyone would have considered the gait of the girl entirely normal. Her mental condition was impaired by the enormous doses of bromide which had been administered in the hope of curing the epilepsy. Recognizing its organic origin, I decided to discontinue the bromides, and, as a result, the epilepsy is no worse and no better; but the girl's mind has considerably improved, and the anæmia, which was aggravated by the administration of the bromides, has been considerably diminished.

The first epileptic attacks associated with a cerebral palsy may occur very soon after the onset of the disease, and may be repeated a number of times during the first week or two. After this a lull sets in, but unfortunately the attacks often recur after several months, sometimes after a year or more, when it is positive that a chronic epilepsy has been developed. According to my own statistics, fully forty-five per cent. of all the infantile cerebral palsies develop epilepsy. This occurs, furthermore, in about fifty per cent. of the cases of hemiplegia, in about thirty per cent. of all forms of diplegia, and in about thirty-six per cent. of patients with paraplegia. I have furthermore observed that the epilepsy does not necessarily develop according to the severity of the palsy, for some of the worst cases of epilepsy I have seen have been observed in cases in which the palsy was extremely slight. The cortical origin of infantile cerebral palsies is sufficient reason for the more frequent development of epilepsy after infantile than after adult cerebral palsies; and if the character of the palsy is such that the lesion in the child can be proved to be capsular rather than cortical, the prognosis as regards epilepsy may be considered favorable. Jacksonian epilepsy is present in some of the palsies, and, if present, can easily be explained on physiological grounds, but, as a matter of fact, it is not observed as commonly as one would suppose, for the simple reason that the majority of patients have been examined after several years have elapsed, and the unilateral order of convulsions has been superseded by a general convulsion. Koenig has given close study to this part of the subject.

Idiocy is developed very often with epilepsy and with some forms of cerebral palsy. Every possible variation of mental deficiency may be present, from slight mental enfeeblement to a complete idiocy. The severer forms are oftener present in the cases of diplegia and paraplegia than in hemiplegia, and naturally enough, for in the former there is a double lesion or disease of both halves of the brain

which will disturb the mental development far more than a unilateral lesion would.

I have found idiocy present in thirty-five per cent. of all diplegias, and in sixty per cent. of all paraplegias, while it occurred in but thirteen per cent. of the subjects of hemiplegia. It was this very high percentage of idiocy in cases of diplegia and paraplegia that forced the conclusion upon me, some years ago, that these congenital paraplegias must be of cerebral rather than of spinal origin. There is no telling in advance whether, in a given case, idiocy will develop or not, for, unfortunately, relatively slight paralysis is sometimes complicated by severe idiocy. Marie claims that there is no disturbance of intelligence in cases of double athetosis. I must insist that there are exceptions even to this rule. The frontal lobes are supposed to be silent parts of the brain, but the defect in intelligence tells a sadder tale of its important functions than the palsy does of the functions of the motor centres.

Various attempts have been made to classify the cerebral palsies of children, and to separate them into distinct clinical groups. The classification according to the period of development, which we have adopted, is, on the whole, the most practical one. The palsies due to intra-uterine disease or to arrest of development, and those due to traumatism during labor, give a distinct history of early onset of all the symptoms. In these children bilateral palsies (diplegia and paraplegia) are more common than in the acute cases, and there is often a very decided defect in mental development. The paralysis is not necessarily complete; at times it is a simple paresis, at other times rigidities and contractures are more prominent than the palsies are. English and American authors have not drawn any sharp clinical lines, as all these forms, although they may vary a little, seem to merge into one another, and are evidently due to similar morbid processes. Little distinguished between rigidities and palsies, and several German authors have adhered to this distinction; but one of the ablest of them, Freud,* has practically adopted the American point of view. Rigidity, contracture, choreic or athetoid movements, paresis, paralysis (unilateral or bilateral), constitute a graduated series of disturbances due to cerebral lesions in children. Each one, or a combination of all of them, may

* This author subdivides the diplegias into four groups: 1. Universal spastic rigidity; 2. paraplegic rigidity; 3. bilateral hemiplegia; 4. bilateral chorea and bilateral athetosis.

be present in a given case, and may vary according to the site and intensity of the lesion. The best proof that all these disturbances of motility are closely related is, that in certain cases of diplegia there is paralysis in the lower extremities and athetosis in one or both upper extremities. Double athetosis is evidently a condition closely analogous to the double hemiplegia, with less paralysis than in other cases. Freud showed that there was a form of acute cerebral palsy with little palsy but distinct chorea; he described this condition as one of choreiform paresis, and recognized an early and a late form. Unilateral and bilateral chorea and athetosis may be developed in the place of a palsy, or the palsy may come first and the chorea and athetosis may represent post hemiplegic disturbances of motion.

The diplegias and paraplegias are generally due to arrest of development or to abnormal birth conditions. (Some distinctly hereditary and family affections allied to these forms will be discussed at the close of this chapter.) Charcot, Marie, Dejerine, van Gehuchten, and Spiller insist that congenital spastic paraplegia may be a spinal affection and due to defective development of the pyramidal tract. For a time Ross adhered to a similar view; but the defect surely involves the brain in a large majority of instances, or else we could not account for the high percentage of idiocy among children with congenital paraplegia. It is in this group of diseases that thoroughly satisfactory post-mortem examinations are needed. One case of Foerster and one of my own have been relied upon. Recently Dejerine has made an interesting contribution to this subject.

Morbid Anatomy.—The study of the morbid lesions underlying these cerebral palsies in children will be facilitated very much by considering separately the three chief groups of cases. A reference to the following table will show that in the prenatal cases large cerebral defects are often found. Porencephaly is present in its widest sense. Half or even more of an entire hemisphere, or of both hemispheres, may be wanting, or a considerable portion of one hemisphere may be poorly developed; in such cases, too, as a rule, the palsy which exists is but one of a long series of symptoms, among which idiocy is by far the most promi-

nent. If it were not for the fact that porencephalic defects occur more frequently in the motor areas of the brain than in any other part, we would scarcely be entitled to associate porencephaly more especially with these cerebral palsies.

CLASSIFICATION OF INFANTILE CEREBRAL PALSIES.

Groups.	Morbid Lesion.
I. Paralyses of intra-uterine onset	Large cerebral defects. (Porencephaly). Defective development of pyramidal tracts (p. 446). Agenesis corticalis. (Highest nerve elements involved.)
II. Birth Palsies	Meningeal hemorrhage, rarely intra-cerebral hemorrhage. Later conditions: Meningo-encephalitis chronica, sclerosis, and cysts; partial atrophies.
III. Acute (acquired) Palsies	Hemorrhage (meningeal, and rarely intra-cerebral); thrombosis (from syphilitic endarteritis and in marantic conditions; embolism. Later conditions: Atrophy, cysts, and sclerosis (diffuse and lobar). Meningitis chronica. Hydrocephalus (seldom the sole cause). Primary encephalitis; polio-encephalitis acuta (Strümpell).

In addition to these large defects in cerebral development, other changes occur in the brain which are not quite as conspicuous, but are fully as effective in the way of producing serious symptoms and serious conditions during life. This condition has been properly designated "agenesis corticalis." It implies a defective development of the cellular elements of the cortical, and particularly of the pyramidal, cells, and is not restricted to any one part of the cortex, but involves all parts of the hemispheres about equally. This condition is present in central amaurotic idiocy, associated with similar changes in the gray matter.

The morbid lesions in *birth palsies* have been satisfactorily cleared up within the past few years. We owe much of this knowledge to the successful demonstration by Dr. Sarah McNutt of a case in which there was wide-spread meningeal hemorrhage in consequence of traumatism during labor. (Fig. 110.) Dr. McNutt furnished positive proof of a condition which Little suspected years ago, in attribut-

ing these palsies to the difficulties during birth. There can be no doubt but that in a vast majority of cerebral birth palsies meningeal hemorrhage, more or less diffuse over one or both hemispheres, is the direct cause of the disease.

The failure to recognize this simple condition has been due to the fact that the cases examined for the purpose of determining the morbid states underlying these palsies have lived for a number of months, or even a year, and at the end of that time the character of the initial lesion cannot be recognized. A very instructive case of this description is the one referred to

FIG. 110.—Meningeal Hemorrhage at Birth. Death on the twenty-second day. (McNutt.)

in a previous chapter (page). The child was born asphyxiated after forty-eight hours continuous labor pains, and an absolutely dry labor at that. From the very first day of its life until the age of six months, when I first saw the child, it had innumerable epileptic spells. The child died at the age of one year. The autopsy and a careful microscopic examination revealed a chronic meningo-encephalitis. This condition was in all probability due to a meningeal hemorrhage, though the traces of the hemorrhage had so thoroughly disappeared that it was difficult to prove this with absolute certainty.

In some instances an original meningeal hemorrhage, particularly if it be more or less circumscribed, may lead to the formation of a limited atrophy, in which case a condition resembling porencephalus may be the result. A few years ago Kundrat described a condition of minute hemorrhages from the meningeal veins, which he considered to be caused by a compression of the longitudinal sinus, in consequence of the displacement of the parietal bones.

Such hemorrhages Kundrat thought were found rather frequently in the brains of healthy children. It is doubtful whether we can connect this form of venous hemorrhage with the subject which we are now considering.

The amount of damage done during labor can be gauged to a certain extent by the symptoms during the first few days of life. If there is wide-spread meningeal hemorrhage convulsions set in at a very early period; the child is apt to be paralyzed at once, and is also in danger of lapsing into coma. Fortunately a number of the severer cases die promptly within a few days, but others again survive, and not a few of these become hopelessly crippled for life. According to the amount of injury done, and according to the special parts of the brain covered by the hemorrhage, the form of the palsy, the degree of mental enfeeblement, and the severity of the epilepsy will vary not a little.

The morbid anatomy of the acute cerebral palsies has given rise to the largest amount of discussion. Hemorrhage, embolism, and thrombosis, the conditions which give rise, in the vast majority of cases, to the apoplexy of the adult, are important factors also in the causation of the acute cerebral palsies of children. This conclusion was forced upon us and others by a careful review of the autopsies made by different authors.

Peterson and myself analyzed the records of seventy-eight autopsies in infantile hemiplegia, as follows:

Lesions.	Number of Cases.
Atrophy, sclerosis, and cysts (terminal conditions)	40
Porencephalus	2
Hemorrhage	23
Embolism	7
Thrombosis	5
Tubercle	1
Total	78

In 35 cases of hemiplegia hemorrhage occurred in 23, embolism in 7, and thrombosis in 5 cases. Professor Osler, who examined 90 brains, found a vascular lesion in 16 only, and among these 16, 7 were due to hemorrhage, and 9 to embolism; but we must not forget that a large number of the cystic conditions may be due to hemorrhage or em-

bolism, and that many a case which is quoted as typical atrophy or porencephalus may also belong to the same category. There has been some little difference of opinion among recent writers on this subject with regard to the relative frequency of these various conditions. Some are in favor of the greater frequency of embolism ; others, like myself, believe that hemorrhages are most commonly the cause of these conditions; while according to Gowers thrombosis of the superficial veins is a very noteworthy initial lesion. The probability is that all these accidents occur, and that

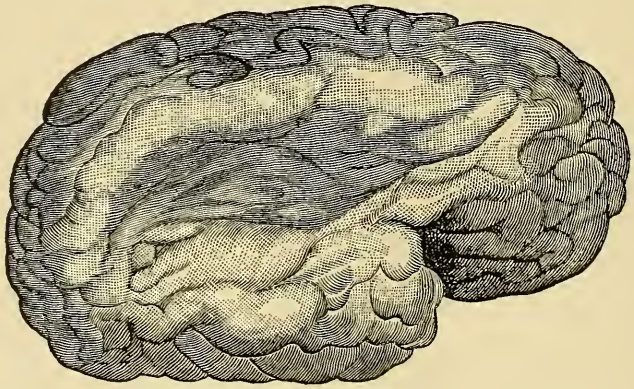


FIG. 111.—Cyst formed by Softening of Brain Substance, secondary to Obstruction of the Middle Cerebral Artery beyond the First Branch (to Inferior Frontal Convolution). The cyst wall has fallen in from escape of its contents. Child nineteen months old. Death seven months after onset of paralysis. (After Ashby and Wright.)

there is no satisfactory reason for claiming more for the one than for the other finding ; but with regard to the occurrence of hemorrhages it is fair to inquire what morbid condition of the blood-vessel leads to the frequent occurrence of hemorrhage in children.

In my endeavors to elucidate this part of the inquiry I have been compelled to fall back upon the fatty degeneration of the blood-vessels which Recklinghausen describes in his masterly Monograph, and which he claims is a not uncommon finding in the brains of children. For the present we can do no better than to adhere to this view of Recklinghausen, and to accept it as a partial explanation, at least, of the frequent occurrence of hemorrhage. Some cases are of course explained by the more delicate structure of the blood-ves-

sels in children as compared with the adult. But there is still another striking difference between these infantile and adult apoplexies. In the adult the majority of hemorrhages occur in the vicinity of the internal capsule. In the child they occur in or near the cortex. Mendel has shown that the greatest pressure is exerted in the branches of the middle cerebral artery, and that any increase of pressure would naturally cause a rupture in one of these. We would have to seek some other explanation for the frequent occurrence of cortical and meningeal hemorrhages, and perhaps they will be sufficiently accounted for by the more delicate structure of these vessels in children. Occasionally intra-cerebral hemorrhages, embolism, and thrombosis occur in

FIG. 112.—Section through Portion of Motor Cortex, removed by Dr. Gerster during an Operation for Localized Epilepsy, associated with Old Left Hemiplegia in a Boy Twelve Years of Age. Specimen was stained according to Van Gieson's method. The pia, P, which is greatly thickened and altered, dips down between two folds of the cortex, C. B, increased number of thickened small arteries; just above, to the right, a large artery with thickened walls. H points to a recent clot. The line, *a b*, denotes the part of cortex examined under higher power and represented in Fig. 146.

children in exactly the same manner in which they occur in the adult, and they give rise to clinical symptoms in nowise different from those of ordinary apoplexy. An interesting case of this sort I had occasion to observe in a boy, two and a half years of age, who after a simple attack of tonsillitis developed right hemiplegia with motor aphasia, without coma and without convulsions. The aphasia disappeared after a few days, and the hemiplegia also improved;

within a period of a few weeks the young boy was entirely well, and scarcely retained a trace of the apoplectic condition. There was no history of syphilis in the case, and the entire development and retrogression of the symptoms reminded one of an adult apoplexy. The possibility of thrombosis could not be excluded, but whether hemorrhage or thrombosis, there can be no doubt about this, that the lesion was intra-cerebral and not cortical, as in the majority of cases in children. Since my own case was published, Dejerine, in 1891, reported three cases in wnich there were distinct hemorrhagic lesions in the vicinity of the larger ganglia.

Heart disease, rheumatism, scarlet fever, and pneumonia, are the conditions which predispose to embolism. Thrombosis may be suspected in cases of children dying of marasmus, but under such conditions the existence of a palsy, is, as a rule, overlooked, and of little practical importance.

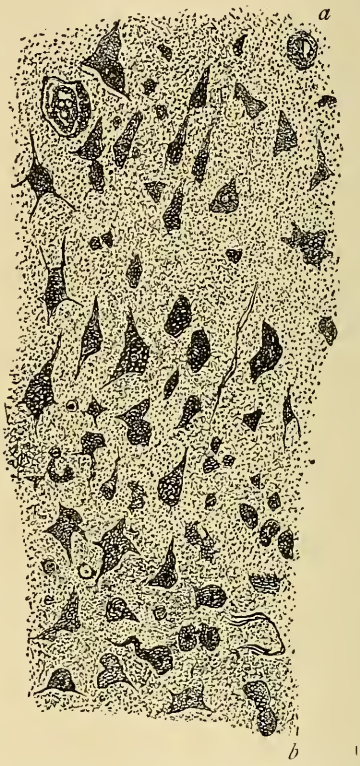


FIG. 113. — Variously Degenerated Cells of the Cortex in the vicinity of the line *a b* of the preceding figure. Near *a*, small blood-vessels, walls thickened. Large pyramidal cells are misshapen, and exhibit granular disintegration. Cells diminished in number. (Drawn from specimen.)

Thrombosis may also be the result of arterial changes due to hereditary syphilis, but I wish distinctly to impress upon the reader that in these cases of cerebral palsies syphilis does surely not play as important a *rôle* as is imputed to it by a few authors, who have generalized from the examination of one or two cases.* Gowers has advanced important reasons for the supposition that thrombosis and occlusion of the middle cerebral veins are of frequent occurrence in children, and that some of the cases of infantile hemiplegia would be more likely to be due to this condition than to embolism resulting from endocarditis. He states, furthermore, that the thrombosis cannot be distinctly demonstrated post-mortem, because the thrombosis is, as a rule, continued into the sinuses, and a sinus thrombosis would be much more striking, and would be apt to conceal the venous thrombosis which led to it.

* I have had no reason to change these views.

Whatever the initial lesion of an acute cerebral palsy may be, if the patient survive a number of years, secondary changes may be set up in the brain which will successfully conceal the initial lesion. Cysts, large areas of softening, atrophy, sclerosis (diffuse and lobar), are a few of the changes frequently observed.

There is no telling in advance of the post-mortem examination, with any degree of certainty, which secondary lesion will be found in the brain of a

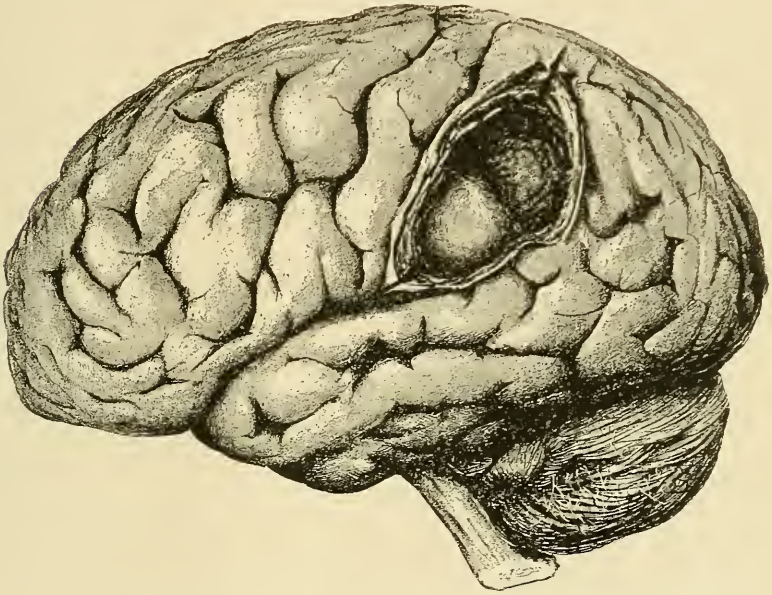


FIG. 114.—An Old Hemorrhagic Cyst. The cyst walls have been cut to expose tumor underneath (compare with Fig. 150). Right hemiplegia at age of six and a half years; death two years later.

child that has been afflicted with an acute cerebral palsy for a number of years. Cysts are so frequent that I have in a number of instances been able to predict the presence of this condition, particularly in those cases in which there was little idiocy associated with the palsy, and in which everything seemed to point to an initial lesion of considerable intensity, but limited in extent. And in several other cases that came under my own notice for operation the surgeon found a cyst in the motor area, which I had predicted to be the probable condition.

Various forms of sclerosis are among the most frequent sequelæ of the initial lesions of infantile cerebral palsies.

This sclerosis evidently starts from a focal region and rapidly speads throughout the brain. If we bear in mind that the hemispheres are traversed by innumerable fibres which are intimately connected with one another, we can understand why a sclerosis should follow upon a relatively small focus of disease, just as secondary degeneration follows upon a lesion in the motor area, and affects all the fibres transmitting centrifugal impulses. This sclerosis, which develops after an initial lesion, is largely responsible for the symptoms which are so frequently associated with these palsies. I refer to idiocy and epilepsy.

In the table given at the beginning of this discussion I have, for the sake of completeness, inserted chronic meningitis as an occasional cause of an acquired cerebral palsy. It is not entirely accurate to attribute a cerebral palsy to a chronic meningitis, but it is better to say that both the chronic meningitis and the cerebral palsy are the result of a cerebro-spinal or of a convexity meningitis which occurred early in life, and which the child survived. These cases are sometimes to be differentiated from others by the persistent paralysis of various cranial nerves in addition to the paralysis of the extremities.

The reader may be surprised to find polio-encephalitis acuta given as the very last morbid condition underlying acute cerebral palsy. Strümpell's idea was a fascinating one indeed, and it is to be regretted that later post-mortem findings did not support his theory. The analogy which this author drew between the infantile spinal paralysis and the infantile cerebral palsies was based upon clinical resemblance, but we well know that symptoms which are scarcely to be differentiated from one another may be due to a variety of morbid processes. Strümpell maintained that there was a close clinical and pathological relation between polio-myelitis and polio-encephalitis acuta. More recently he has modified his views, and now claims that acute encephalitis of the gray, as well as of the white matter, may constitute the basis for these acute cerebral palsies in children. The actual proof which Strümpell offered for his views was based upon the occurrence of two cases in one family, reported by Moebius, in which two children were affected at one and the same time; the one with a typical polio-myelitis, and the other with a spastic cerebral palsy without atrophy. This may have been a mere coincidence, and further evidence must be forthcoming before we can accept this as proof of the theory. Strümpell himself refers to two cases in the adult in which the diagnosis of hemorrhage was made during life, and in which, at the post-mortem examination, acute encephalitis was found. From this we may infer the possibility of a similar occurrence in children, but until such a condition can be satisfactorily established there is no good reason to accept encephalitis as more than the rarest of all causes that give rise to infantile cerebral palsies. A case of (supposed infectious) encephalitis of the island of

Reil, reported by Felsh, is not convincing. Lesions affecting the motor areas give rise to a palsy; similar lesions may affect silent parts of the brain, and the result of such lesions may be a general cerebral disturbance, defective intelligence, or chronic epilepsy. The fact has been well emphasized by Freud who speaks of cases of "cerebral palsy without palsy."

DIFFERENTIAL DIAGNOSIS.—Infantile cerebral palsies are frequently confounded with the acute spinal affections. The cerebral cases are characterized by the hemiplegic, diplegic, or paraplegic form of paralysis, by spastic rigidities and contractures, by increase of the deep reflexes, by the entire absence of any considerable degree of atrophy, and by entirely normal electrical reactions. Anyone who is able to recognize the difference between lesions in the first and second divisions of the motor tract will be able to distinguish between the cerebral and spinal forms of infantile palsies. Difficulties may, however, arise in the cases of mild types of cerebral palsy in which the spasticity and contractures may be but slightly developed, and the reflexes may not be very much increased; but even under such circumstances the hemiplegic distribution of the palsy and the entire absence of all changes in the electrical reactions will help to demonstrate the cerebral character of the disease. In other patients, the presence of athetoid or choreic movements in one half of the body, with slight exaggeration of the reflexes, will indicate a former cerebral palsy, the paralysis having wellnigh disappeared. Spinal and cerebral palsies may occur at different periods of a child's life, but I have seen only one instance of this among the large number of cases that have come under my notice. The spastic paraplegia due to traumatism during labor is to be differentiated from hereditary spastic paralysis (see Chapter XXI.) and from syphilis of the spinal cord (see Chapter XVII).

Although the diagnosis is simple enough, the knowledge of these infantile cerebral palsies is still so very imperfect among physicians that cases coming under this category are frequently overlooked. I would particularly urge the examination of patients with peculiar forms of athetosis or chorea in order to determine whether these are not associated with some traces of a preceding cerebral

palsy. If such children exhibit hemiparesis with slight defects in mental development, or with occasional epileptic attacks, the true nature of the disease can readily be discerned.

PROGNOSIS.—In the congenital forms of cerebral palsy the prospects as regards life and the normal development of the child cannot be formulated until after a period of some weeks or months. It is a fortunate circumstance on the whole that children who come into the world with a severe cerebral lesion succumb to it at a very early period, but of those who survive a fair proportion develop idiocy as well as epilepsy, and are frequently permanently crippled. If convulsions occur during the earlier weeks of life the severity of the lesion may be inferred from this fact. If such a child, moreover, show no mental awakening, the probability of more or less complete idiocy is very great indeed. If, after a few weeks or months, the convulsions are noticeably diminished, if the child shows any tendency to a tolerably normal use of its legs, and if it begins to take notice of its surroundings, a more favorable prognosis may be given. As long as contractures do not develop the child may acquire a fair use of its extremities. Diplegia and paraplegia are more apt to be associated with cerebral deficiency and epilepsy than is unilateral palsy.

In the acute cases there is much uncertainty for some time after the onset of the attack, and no definite statement is warranted regarding the permanency of the paralysis, or the mental condition of the child until several weeks have elapsed, when it will be seen whether there is any tendency or not to recovery. If a child with an acute form of infantile hemiplegia shows some improvement after a few weeks, either in the leg or in the arm, and if the speech that was lost begins to return, there is every reason to be hopeful regarding the ultimate outcome of the disease ; but if weeks and months pass without any such favorable change the probability of permanent crippling of the mind and body is very great. It is, however, a matter of common experience that the child may do extremely well for a number of months, for a year, or even longer, after an attack, when convulsions may reappear and the epileptic habit may be-

come established. I have paid considerable attention to this point, and find that among the cases examined by me epilepsy occurred in fully forty-five per cent. of all the cerebral palsies, and nearly one-half of the cases of cerebral hemiplegia developed epilepsy at a later period. It would seem from this that the unilateral brain lesion leads to epilepsy more frequently than the double lesion does, for according to the same statistics epilepsy is present in about twenty-nine per cent. of the diplegias, and in about thirty-six per cent. of the cases of paraplegia. My own results regarding the development of epilepsy are corroborated by the statistics of Gaudard, Osler, and Wallenberg. The fact that fewer cases of diplegia and paraplegia survive may account for this apparent difference in favor of the double lesions. If a child that has passed through an apoplectic seizure, and has some form of congenital paralysis is taken with convulsions, it is more than probable that such convulsions are the first of a series that may lead to the development of chronic epilepsy, and from this time on the prognosis becomes extremely grave.

TREATMENT.—The treatment of infantile cerebral palsies is not altogether devoid of interest. These diseases call for deliberate non-interference at the start, and for sober judgment in therapeutic matters during the later stages. The treatment of the later stages will be practically the same whether a case be congenital or acquired. If a child but a few days old exhibits a tendency to drowsiness or to convulsions its brain has in all probability sustained a serious injury during labor. It should be kept quietly in its crib or bed, and the greatest attention should be paid to its nutrition. These children are often unable to swallow and unable to suckle at the breast, and for this reason must be carefully fed with a spoon. Milk, properly prepared, according to the age of the child, is the very best nourishment. If there is a tendency to convulsions, or if marked convulsive seizures occur, the child may be given very minute doses of bromides, or minimum doses of morphia or chloral. A drop of the solution of Majendie, or a grain of chloral per rectum, will be quite sufficient in the case of a new-born babe. If convulsions continue in spite

of these measures, inhalations of chloroform may be practised, however young the child may be, but of course with the greatest possible care. All other measures, such as counter-irritation, mustard-baths, and the like, are practically useless.

In the initial stages of an acute cerebral palsy the same measures should be employed which the physician would resort to in the case of an adult apoplexy, allowing always for the difference in the ages of the patients. Absolute rest is the first condition of treatment. An ice-bag may be applied to the nape of the neck, or to the head, and if it do no good, it will at least do little harm. The use of ergot or of the nitrite of amyl, as has been advocated in adult apoplexies, I cannot favor. The fewer drugs that are administered the better it will be, and the physician will do well to limit himself entirely to the administration of calomel in doses sufficient to procure free purging of the bowels, and small doses of bromide, which may be given in order to secure rest for the disturbed brain. At a later period the bromides may be combined to advantage with the iodides, but in every case in which the digestion or the general nutrition suffers from the administration of these drugs, their use should be prohibited for the time being; for the good that they can effect will not counterbalance the evils that follow upon a gastritis and its attendant malnutrition.

As soon as the symptoms of the initial period have passed, the physician is compelled to prescribe some form of treatment for the paralysis and the other symptoms of the disease. The paralysis is the natural result of the lesion, and cannot of course be removed by any therapeutic measures. It is well to explain this distinctly to the parents and relatives, and to state at once the chronic nature of these troubles, and the length of time that will probably elapse before any great change can be expected to take place. As soon as the active period of the disease has passed, massage and electricity should be applied to the paralyzed parts, but let the brain severely alone. The effect of electricity upon the circulation within the skull is altogether too uncertain to justify us in tampering with it in any way. Electricity applied to the extremities can do no

harm, and serves an excellent purpose as a means of exercising parts that cannot be moved by the will. As these parts respond to the faradic as well as to the galvanic current, I am in the habit of using the former chiefly, and of using a current strong enough to produce mild contractions. Powerful currents should be avoided. The electricity should be administered in sittings from ten to fifteen minutes every day, or every other day, and should be given by the physician, or by an extremely competent nurse under the guidance of the physician. If passive movements are combined with ordinary friction the paralyzed parts will be kept in a good state of nutrition, and the tendency to contracture may be overcome.

As in the cases of spinal paralysis, so in these cerebral palsies, orthopedic measures should be resorted to as soon as contractures have become permanently established. I have seen many a child walk about fairly well a few weeks after the orthopedic surgeon has been allowed to cut the tendons, and to provide the proper orthopedic appliances. Contractures, if once formed, are rarely, if ever, recovered from in spontaneous fashion, and if it can be proved that the contractures are the chief hindrance to the child's progress in walking, full liberty should be given to the orthopedic surgeon; Eulenburg, Hoffa, and others have obtained good results by tendon-transplantation. I have been especially pleased with what Dr. Gibney has been able to do in several cases of marked disturbances of the upper extremities that have occurred in cases of infantile cerebral palsy. I refer in particular to the persistent athetoid-choreic movements of the upper extremities so common in these cases. By the application of some simple form of restraint such post-paralytic movements may be successfully inhibited. In the case of a young man, seventeen years of age, who had acquired a cerebral palsy when he was ten months old, and who had exhibited very annoying athetoid movements of the upper extremities from that time on, the simple splint arrangement which prevented the possibility of these movements, and which was worn for a number of months, was sufficient to inhibit the athetoid movements altogether; and in still another case a marked post-hemiplegic chorea was much improved, if not altogether inhibited, by placing the arm in a plaster-of-Paris bandage for a period of two months.

The treatment of the idiocy must be conducted on the same lines as in other forms of idiocy.

By far the most important task is the treatment of the epilepsy associated with the palsy. We have stated elsewhere that this epilepsy is due to the secondary changes in the brain, which have developed as a natural result of the

original lesion which gave rise to the palsy. In all these cases the epilepsy is practically a focal epilepsy, although its clinical manifestations are not often of the strictly Jacksonian order. It was quite natural that Horsley's suggestions with regard to the proper treatment of focal epilepsy should be applied to these forms of epilepsy with infantile palsies.

Horsley himself has reported upon two such patients, and in both of these a cystic condition was found. Freud reports a case of Oppenheim's in which a meningeal cyst over an arm centre was evacuated, but the brain itself was not operated on by the surgeon; the athetosis and epilepsy in these subjects were considerably diminished. The author has had several cases of cerebral palsy with epilepsy operated upon. The first case, a boy, six years of age, had acquired right hemiplegia at the age of five, and soon thereafter developed convulsions, which invariably began in the right hand. This right hand also exhibited athetoid movements. Dr. Gerster trephined over the motor centre for the right arm, and exposed the greater part of the arm and leg centre in the left half of the brain. The boy did well for three months after the operation; his attacks disappeared altogether, but the ultimate result cannot be stated, as I lost sight of the patient.

Another case was that of a girl, sixteen years of age, with right hemiplegia and epilepsy. Wyeth performed the first operation on this patient, exposing the dura over the entire motor area, and a very large portion of the cortex was inspected after the dura had been incised. The exposed cortical tissue was tested by the faradic current, and responded normally to mild currents; as the cortex seemed to be normal I did not urge its excision. I regretted at that time not having urged the complete excision of the arm centre. I do not, however, consider Horsley's reason for removing such centres entirely valid, since secondary changes, which have been established, seem, after all, to be the main source of irritation, and if one focus of disease be removed I am confident that other parts of the hemispheres will soon attain to the dignity of epileptogenous centres. It is, furthermore, well established at the present day that convulsive seizures may result from irritation of almost any part of the cortex. In this case there was very little improvement excepting that the epileptic attacks were transferred after the operation to the other half of the body, whose centres had not yet been exposed. I have had the other half of the brain exposed more recently by Dr. Gerster, but for reasons which I have suggested did not urge the excision of any of these parts, and I fear that the girl will remain an incurable epileptic.

A third case was one operated on by Dr. Wyeth. The patient was a man, thirty-two years of age, who had had an apopletic attack at the age of two years. As a result of this he exhibited a left hemiplegia with considerable atrophy, with contractures of the left upper extremities and athetoid movements of the left hand; furthermore, pes equinus of the left side with a

tendency to equino varus. I made the diagnosis of an old infantile spastic hemiplegia, probably due to meningeal hemorrhage over the arm centre in the right hemisphere. Dr. Wyeth exposed this centre according to my directions, and at this point was found a distinct hemorrhagic discoloration of the pia, with the development of adhesions between the pia and the cortex. I advised that these adhesions be incised, but did not urge removal of the cortical tissue, though in this case I am free to say, since the arm was already paralyzed and practically useless, a very liberal excision of this arm centre might well have been practised. The man recovered well from the operation, and his convulsive seizures have been very much diminished during a period of nearly three years. In October, 1894, Dr. Gerster operated upon a fourth case (infantile hemiplegia with epilepsy) in a boy, aged twelve; the epileptic fit began in the left (paralyzed) arm, and then became general. The greater portion of the arm centre was excised, showing the effects of old hemorrhage (Fig. 145); the attacks have not been sensibly diminished. Similar cases have been reported by Weir, Keen, Roswell Park, Angell, and Starr. In Weir's case a large cyst was found, which was carefully drained, but the child, four years of age, died five hours after the operation. In one of Keen's cases the child died twenty minutes after the operation, and the brain exhibited a very considerable atrophy of the entire left hemisphere. The greater danger of all these operations in young children should be taken into account, and for this reason I now hesitate to have the operation performed upon any child under the age of four years. Wildermuth has reported two cases. In the one case he removed atrophic cortical tissue, and evacuated a cyst underlying it. Three years after the operation the patient was said to have been improved. In another case he also performed excision of an atrophic portion of the cortex, and the attacks were said to have been inhibited up to ten months after the operation.

Dr. Angell, of Rochester, has reported upon a case of infantile hemiplegia (birth case) with imbecility and epilepsy. A simple craniotomy was done, exposing a cyst in front of and above the ascending frontal region of the left side; a slight improvement followed. If a cyst is as clearly limited as in this case, removal of the cyst, or at least drainage of the same, would have been a more rational and radical operation; the improvement following upon craniotomy can be explained as the result of a release of pressure.*

The results of the last few years have been much more encouraging (cases of Starr, Chipault, Krause, Oppenheim, Sonnenburg and my own); if all the symptoms of a case point clearly to an initial focus of disease, surgical procedures are quite in order. As cysts are extremely frequent in these infantile cerebral palsies, the evacuation and removal of such cysts will often be followed by considerable relief. I believe that it is due to the frequency of such cysts that relief follows so many operations for epilepsy.

If surgical procedures are unavailing, the ordinary anti-

* While this edition was passing through the press, Cushing reported the successful removal of large meningeal clots a few days after birth! "The procedure itself, though delicate in performance, need not entail serious consequences."

epileptic measures may be employed. Bromides and chloral should be given in moderate doses, but there is little use in pushing these drugs to the extreme, for we cannot expect to cure an epilepsy that is dependent upon an organic brain lesion.

It will be best in this connection to study a disease which has evident relations to other family affections (see Chapter XXI). Although it is a cerebro-spinal disorder, the author's studies on this disease were made largely in connection with the study of the infantile cerebral palsies.

AMAUROTIC FAMILY IDIOCY.

This name was given by the author to a disease not altogether rare,* affecting several members of one family, and characterized by an impairment of all the mental functions, by a progressive weakness of some or of all the muscles of the body, and by a defect in vision (associated with changes in the macula lutea and subsequent optic nerve atrophy), terminating in complete blindness. The disease is fatal, the children dying, as a rule, in a condition of complete marasmus before the end of the second year of life.

History of the Disease.—In 1881 Waren Tay described "symmetrical changes in the region of the yellow spot in each eye of an infant." He had noted the fact that the child's cerebral development was slow, and also that the optic discs were apparently healthy at first, but that, in the region of the yellow spot, there was a conspicuous, tolerably large white spot more or less circular in outline, and showing at its centre a brownish-red, fairly circular spot contrasting strongly with the white spot surrounding it. Tay likened this "cherry-red spot" to the appearances which are well known in cases of embolism of the central artery of the retina, and believed the changes to be "possibly congenital." Subsequently he noticed a gradually developing atrophy of the optic discs, while the changes in the macula lutea remained the same as before. Tay also reported that three similar cases had occurred in this family. These same ophthalmoscopic findings had been noted by a number of oculists in Europe and in America.

So long ago as 1887, without any knowledge of the cases described by the oculists, the author reported what appeared to be a peculiar form of idiocy associated with blindness. Some years later I became aware of the family character of this affection, and it was then shown by Kingdon, of Nottingham, that the cases reported by the oculists and those reported by me were identical. But it was not until the year 1898, after a much larger experience with this category of cases, that I was convinced that this condition deserved to be considered as a clinical entity, and in that year I proposed the name Amaurotic Family Idiocy, which has been universally adopted.

* In 1904 Heveroch had collected the records of 86 cases, of which 44 occurred in America.

Risien Russell has been the only one to use the designation "Infantile Cerebral Degeneration." The name is wholly inadequate, and in view of our present knowledge of the pathology of the disease, does not do justice even to the morbid anatomy of Amaurotic Idiocy. The symptoms as described by me have been endorsed by all subsequent writers. Among these I wish especially to mention Peterson, Hirsch, Koplik, Kingdon, Russell, Falkenheim, Higier, Schaffer, Fry, Spiller, McKee, Mary Buchanan, and Shumway.

FIG. 115.—Case of Amaurotic Family Idiocy, 14 months old. Patient of Dr. Cotton, of Chicago, showing well-nourished condition of child in earlier stage of disease.

SYMPTOMATOLOGY. — The children are born healthy (several of them have been unusually robust) and do well until the age of four, five or six months. At about that time they become listless, apathetic, move their limbs very little, and show the first signs of impaired vision. The child is unable to hold its head up straight or to sit up, and as time goes on, instead of gaining strength in the use of its muscles, its limbs become limp, in some instances spastic and more or less paralyzed. The reflexes may remain normal, at times they may be a trifle subnormal, and in the spastic cases they become exaggerated. An unusual sensitiveness to sound (hyperacusis) is observed at a very early date, so that the child is startled by the slightest noise, by a clapping of the hands, etc. Oppenheim speaks of this symptom as an increased motor acoustic reflex. Convulsions may occur, but are not an integral feature of the disease. After the lapse of a few months the mental impairment is distinctly increased and blindness becomes absolute, and the child lapses into a condition of extreme marasmus, in which it dies, as a rule, before the end of the second year (Fig. 115a).

We may sum up the chief symptoms as follows: (1) A mental impairment observed during the earlier months of life and leading to absolute idiocy. (2) A paresis or paralysis of the greater part of the body which may be either flaccid or spastic. (3) The reflexes may be normal, deficient, or increased. (4) A diminution of vision terminating in absolute blindness (cherry-red spot

in the region of the macula lutea with subsequent optic nerve atrophy). (5) Marasmus, and a fatal termination, as a rule, at about the age of two years. (6) Unusual sensitiveness to sound (hyperacusis). (7) The appearance of the affection in several members of the same family.

In some of the children I have noted nystagmus and strabismus. A few other symptoms in addition to those described above have been mentioned by Falkenheim. These are "explosive laughter" and "disturbances of deglutition." The former I have not observed, but it is very probable that it may occur in some of the cases, and the latter may be taken to be an accompaniment of the great mental and physical deterioration; and the occurrence of such bulbar symptoms need not astonish one, in view of our present knowledge of the anatomy of the disease.

FIG. 115a.—Child in Last Stages of Amaurotic Family Idiocy, showing extreme emaciation and (unusual) contractures. Author's case.

The ocular symptoms constitute a most striking feature of the disease. In every instance in which the opportunity for ophthalmoscopic examination has been given, this peculiar condition was found present, but I wish to state distinctly that the disease can be diagnosticated, as has been done, before the occurrence of the typical changes in the region of the macula lutea. In Higier's case optic nerve atrophy was much more pronounced than the changes in the macula lutea. All writers on the subject have fully appreciated the characteristic symptoms of Amaurotic Idiocy, but the importance of the rapidly developing marasmus has not been sufficiently insisted upon by many of them. I hold it to be one of the important symptoms of the condition.

ETIOLOGY.—The causes underlying this disease are still somewhat obscure. Blood relationship between the parents has been noted in some instances and a marked psycho-neurotic taint in others. In a few instances the mother has sustained some injury during pregnancy. The family predisposition is made evident by the fact that twenty-eight cases which came under my notice have occurred in fifteen families. Carter was the first to call attention to the fact that all cases reported have occurred among Hebrews, and up to the present time not a single entirely satisfactory case has been reported as occurring among other races. It is truly astounding

that this disease should be thus limited, because other diseases to which it is more or less closely allied have been observed among all races and all nationalities.

It is well to state explicitly that this is not a syphilitic disease, and this point should be insisted upon, because hereditary optic nerve atrophy occurring later in life has been shown to be due frequently to hereditary syphilis, and there are other family affections associated with dementia which are due to this same cause.

DIAGNOSIS.—There can be little difficulty in diagnosticating this form of disease if the symptoms as enumerated above are kept in mind. Hereditary spastic diplegia, which sometimes occurs in families, may give rise to some confusion, but in this disease the entire absence of visual symptoms, the lack of mental impairment, the marked spasticity of all limbs, and the fact that the children show no tendency to marasmus will help to bring out the true diagnosis. Hereditary optic atrophy may also be confounded with Amaurotic Idiocy, but the former only occurs in older persons. Several times the author has been asked to see cases which were supposed to be instances of Amaurotic Family Idiocy which presented the symptoms due to a preceding meningitis; and on the other hand, the author is also aware that in not a few instances the diagnosis of meningitis has been made when the case was a very typical one of Amaurotic Family Idiocy. Rayner D. Batten has reported symmetrical disease of the macula lutea in two brothers beginning at the age of 14 years, and F. E. Batten describes "cerebral degeneration" with symmetrical changes in the macula lutea in two members of a family. These cases are, however, very distinct from the type here described. In Rayner D. Batten's cases there was hereditary syphilis; yet it is of interest to note the family occurrence of ocular changes in and around the macula lutea.

FIG. 116.

PATHOLOGICAL ANATOMY.—In three brains which the author has had an opportunity of examining there were peculiar abnormalities of fissura-

tion (confluence of the fissure of Rolando and fissure of Sylvius and in two instances exposure of the island of Reil). Such abnormalities are generally conceded to be a sign of deficient cerebral development (Fig. 116). In the earlier publications on this subject I spoke of the morbid condition as an Agenesis corticalis, a defect in the development of the highest nerve elements of the cortex. There was also some degeneration of the white fibres in the hemispheres and of the pyramidal tracts in the spinal cord. This degeneration of the white fibres could be traced through the course of the pyramidal tracts in the internal capsule, crusta, pons and medulla. But more striking than

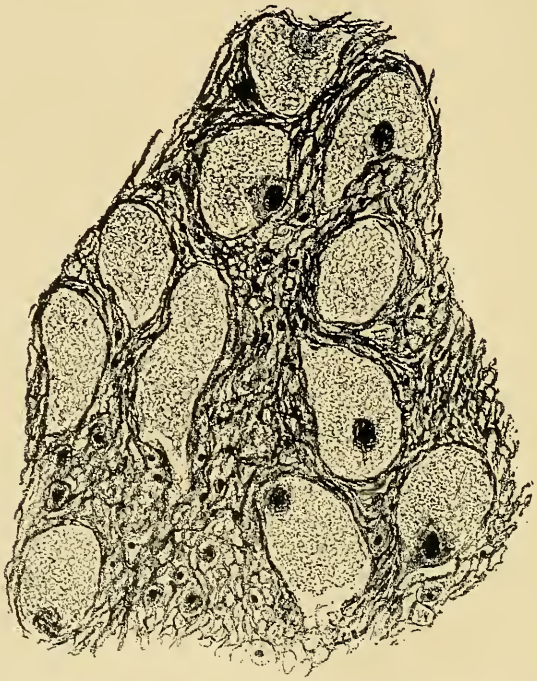


FIG. 117.—Section through XII nucleus in medulla oblongata, showing advanced changes in ganglion cells. Hematoxylin-eosin stain.

this change in the white fibres is the change in the gray matter not only of the cortex, but throughout the entire central nervous system. The changes are found to be the same in the cortex of the brain, in the cranial nerve nuclei, and in the anterior and posterior gray matter from the cervical to the lumbar and sacral segments of the cord. Even the spinal ganglia showed similar changes. Our knowledge of the spinal changes we owe chiefly to Dr. Hirsch, whose findings I have been able to corroborate in a case examined by me only a few years ago. There can no longer be any doubt that the disease is due to a degeneration of the ganglion cells throughout the entire central nervous system. In the cortex, as well as in the gray matter of

the cord, there is scarcely a normal ganglion cell to be seen. As will be noted from the illustrations herewith presented (see Figs. 117–119), the

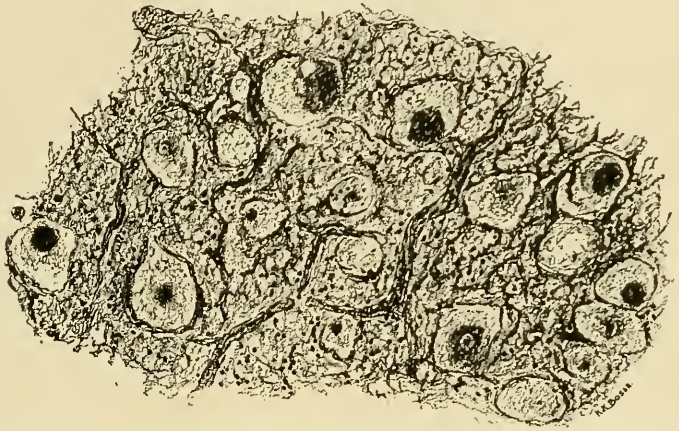


FIG. 118.—Section through cervical enlargement, showing cell changes in anterior gray matter. (Van Gieson stain.)

cell-body is completely altered; the entire cell protoplasm has become disintegrated, leaving a more or less homogeneous mass; the nucleus has been

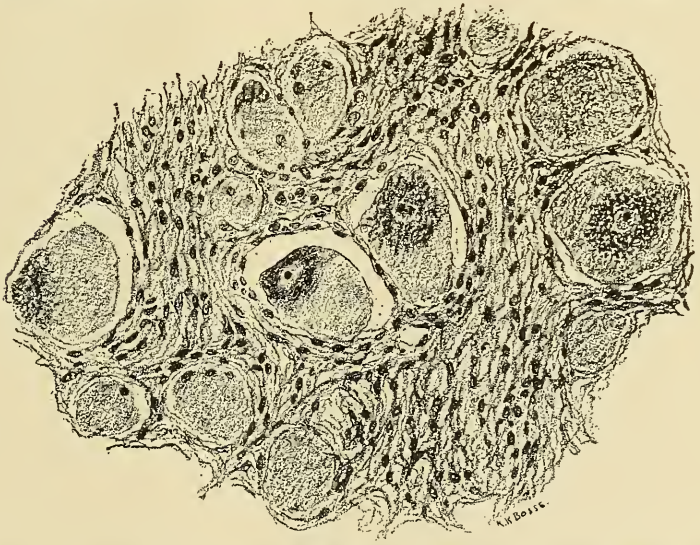


FIG. 119.—Section through a lumbar ganglion. Nissl stain showing marked cell changes.

shifted commonly to some part of the periphery of the cell, and instead of being distinctly differentiated from the rest of the cell-body, it generally

shades off by degrees into the disintegrated structure of the cell. The nucleus is often wanting entirely, and in some instances the whole nucleus appears to have dropped out. The disintegration of the cell-body is so complete that in many specimens the contour alone enables one to infer that these round bodies are the remnants of ganglionic cells. These metamorphosed ganglion cells are frequently surrounded by distinctly pericellular spaces, and are relatively and absolutely so much enlarged that they give a very striking appearance to the cross-section.

Holden, as well as Shumway and Mary Buchanan, have made careful histological examination of the eyes in Amaurotic Family Idiocy. All these writers are agreed that the essential changes in the eyes are "a degeneration of the ganglion-cells of the retina and of the nerve-fibres of the optic nerves and tracts, which are genetically a portion of the central nervous system." They also believe that the white area in the fundus is the result "of the swollen and degenerated ganglion-cells which are present in much greater numbers in the macula region than elsewhere," and do not consider this white area to be due to an œdema of the tissue as was first supposed.

Hirsch, Spiller, Shaffer, of Budapest, and the author have proved that Amaurotic Family Idiocy is due to a degeneration of the entire gray matter of the brain and spinal cord. The degeneration of the white fibres of the anterior and lateral pyramidal tracts is in all probability secondary, but not nearly so marked as one would expect with such advanced disease of the ganglionic elements. It is probable that the relative involvement of the gray and white matter may vary much in different subjects affected with this disease.

Hirsch and some others have supposed that Amaurotic Family Idiocy was an acquired disease; that the morbid changes in the cells were of a distinctly degenerative character, and that these changes were similar to those produced by toxic agents of various sorts. Hirsch went to the extent of claiming that the toxic agent might possibly be conveyed to the child through the mother's milk. It is difficult to explain any family disease on the supposition that it is of toxic origin, and in this instance the author must repudiate this view for the simple reason that several of his patients have not been nursed by their own mother; and it is very evident that, although the symptoms of the disease are not always noticed before the age of three or four months, the development of the disease is due to conditions antedating the time of birth. The author still firmly believes that Amauratic Family Idiocy is due to an arrest of development, and that this arrest of development is followed by a degeneration. Accepting Gowers' and Edinger's views of the inherent or deficient capacity for normal development, we may simply argue that children who are afflicted with Amaurotic Family Idiocy have a very limited capacity for normal development; that their central nervous system is not equal to the functions that they are expected to perform for more than the first three or four months of life, and from that time on a rapid deterioration of all functions is established.

PROGNOSIS.—The disease is invariably fatal; between the age of twelve and eighteen months the children, even though they have been in tolerably robust health at birth, pass into a condition of extreme marasmus. Only one

case in which the diagnosis of Amaurotic Family Idiocy could be accepted has lived beyond the age of two years.

TREATMENT.—No form of treatment has been of any avail. It is very positive that the disease is not due to hereditary syphilis, and for that reason even the ordinary iodide treatment cannot be urged. In several families in which children with this disease have been observed, others have been born and have remained entirely healthy. No further warning should therefore be given to parents of such children; let them, however, exercise special care with reference to the physical development and early training of their offspring.

Freud has reported several interesting cases under the title of "Family Forms of Cerebral Diplegia." Two of the children were bright at the ages of six and five years respectively, but they presented from earliest childhood the following symptoms: Nystagmus, atrophy of the optic nerve, convergent strabismus, awkwardness in speech and in the use of the arms; tremor of the arms and a spastic paraplegia of the lower extremities; no convulsions. In these boys the disease is evidently present in its lightest and most favorable form. The improvement in vision as the one child grew older seems evidence of this, but the resemblance of these forms to those described by me is shown by a third child that died at the age of ten months; it was paralyzed from birth, and idiotic; whether it was blind also is not stated. The father thought the child had died of rickets (more probably of marasmus).

Pelizaeus has described a family disease of which the chief symptoms were: Nystagmus, mental imbecility, disturbances of speech (Bradylalia), awkwardness of hands, spastic paraplegia of the lower extremities. The disease attacked male members of several generations, but was transmitted through healthy mothers. All of those who were attacked showed some symptoms in childhood, and several died young. Pelizaeus was inclined to the diagnosis of multiple sclerosis; but his cases evidently constitute a late form of hereditary spastic paralysis.

CHAPTER XXVI.

TUMORS OF THE BRAIN AND ITS MENINGES.

NEW-GROWTHS within the skull are very common in childhood. There is no period of early life that is entirely exempt from them. Careful studies of cerebral tumors have been made by Steffen, Bernhardt, Starr, Knapp, Bruns, and Oppenheim. The following tables are taken from an article by Peterson:

TABLE I.

Form of Tumor.	No. of Cases.
Tubercle	166
Glioma	42
Sarcoma	37
Cyst	35
Carcinoma	11
Gliosarcoma	5
Angiosarcoma	1
Myxosarcoma	1
Papillary epithelioma	1
Gumma	1
Not stated	35
Total	335

TABLE II.

Site of Tumor.	No. of Cases.
Cerebellum	105
Pons Varolii	42
Centrum ovale	41
Basal ganglia and lateral ventricles	30
Corpora quadrigemina and crura cerebri	25
Cortex cerebri	23
Medulla oblongata	7
Fourth ventricle	6
Base of brain	8
Total	287

From these tables we can infer, first of all, that tubercle is much more frequent than any other form of intra-cranial neoplasm, very nearly one-half of all the cases being of this character. Glioma and sarcoma are the next most common forms, and as the cerebral tissue in earlier years is subject to proliferation, the frequent occurrence of the former need not be a matter of surprise. Cysts differ from tumor in this respect that they are secondary products of some preceding morbid process, very often resulting from an old hemorrhage, or from embolic softening. The contents of a cyst may remain stationary for a very long time, and under such circumstances the symptoms will not be very like those of a growing tumor. In other instances the contents of the cyst suddenly increase and all the symptoms may be those of a rapidly increasing neoplasm.

As for the location of tumors in general, it will be seen that a larger number are found in the cerebellum than in any other part of the brain. The pons and the centrum ovale are next in order of frequency, then come the basal ganglia, and last the cortex of the brain, and the region of the corpora quadrigemina. The medulla oblongata and the base of the brain are rarely the seat of tumor.

ETIOLOGY.—The actual cause of cerebral tumors in children is a matter of considerable speculation. It is only with regard to gliomata that we can state that the natural tendency of the infantile brain to proliferation of its tissue upon the slightest provocation accounts for the frequent occurrence of this special form of neoplasm. Tubercle, sarcoma, and carcinoma of the brain, are always secondary to deposits of a similar character in other parts of the body. Carcinoma is rare in children, and carcinomatous deposits in the brain are correspondingly infrequent, but under exceptional conditions these may occur early in life. A few cases have been observed which were due to an invasion of the growth from the orbit. The exact relation of external injuries to new-growths has been often in dispute. It is more than probable that the accident to which the tumor is referred is, as a rule, merely a landmark in the child's history.

In the fewest instances only can an actual causal relation between the two be established. The hyperæmic condition or the slight hemorrhages which occur at the seat of injury may at times be sufficient to become the starting-point of a latent morbid process. Boys are more liable to brain tumors than girls, and those who are not too careful in utilizing statistical evidence will claim that this fact is to be explained by the greater frequency of injuries to the boys' skulls.

SYMPTOMS.—As in the adult, so in the child, the symptoms of intra-cranial tumor may be divided into two distinct groups. The first includes the general symptoms resulting from increased pressure, and the second group includes those due entirely to the location of the tumor. It is a matter of course that slowly progressive tumors will exhibit less symptoms than those which invade the brain rapidly and soon attain to a very considerable size. A tumor of small size growing rapidly, will produce far more symptoms than one of huge size which has taken years to develop. The symptoms will also vary according to the blood-supply of the tumor, and according to the increase in the amount of hydrocephalic fluid. The general symptoms which we must consider somewhat in detail are headache, nausea, vomiting, stupor, insomnia, convulsions, slow pulse, and double optic neuritis.

Headache is in many cases the most striking symptom of tumor of the brain. The nearer the tumor is to the cortex, and the more it involves the meninges, the more intense on the whole the headaches are likely to be; but I have seen headaches as intense as those of cortical tumors in cases in which the growth was in the vicinity of the ganglia, or even at the base of the brain. In such patients the increase in cerebro-spinal fluid has much to do, in all probability, with the development of the headaches. The intense "boring," "gnawing" pain is generally referred to that part of the head which is nearest to the new-growth; but there are exceptions to this rule as to every other. In children whose fontanelles are not completely closed the tumors may attain to a considerable size without causing intense headaches. These headaches are often the cause of sleeplessness, and both the pain and the insomnia are largely responsible for the rapid emaciation of the child. In some few instances insomnia is present without pain, and is the result of cerebral excitement.

The association of nausea and vomiting with headaches in a child should lead to the suspicion of tumor, particularly if all these symptoms persist for some considerable period of time. The vomiting is characterized by its suddenness, and by its projectile character. Nausea may be associated with it, but in many cases there is no warning given of the vomiting, and it comes as a surprise upon the child, and often occurs entirely independently of meals; at other times during meals. Vertigo is not infrequent in cases of tumor, and is very apt to occur with every change in position of the head. It is very frequent indeed in connection with tumors of the cerebellum, or of the pons, which is explained by the relation of these parts to the auditory nerve.

Convulsions in children are so common that they alone need not give rise to the suspicion of tumor, but if associated with constant headaches, with

vomiting, and if all these symptoms are frequently repeated, the suspicion may be well founded. These convulsions are very often general, but in addition to the general convulsions, or without the occurrence of such, localized convulsions may occur if the neoplasm is in some part of the motor area. Such localized convulsions, associated with headaches, vomiting, and with optic neuritis, point very definitely to an intra-cranial growth.

Double optic neuritis and the complete optic atrophy following it are important symptoms of intra-cranial tumor. It is certain that there is no symptom which if present, is more valuable than an optic neuritis; but, on the other hand, tumors may attain considerable size without the development of optic neuritis. This neuritis is in all probability due to increase of intra-cranial pressure. Other theories which have been proposed do not explain all the peculiarities of optic neuritis as well as the theory of increased pressure does. The fundus should be examined in every doubtful case of brain tumor, for normal vision is not incompatible with a considerable degree of optic neuritis, and changes in the optic nerve may occur, and do occur, with tumor in any part of the brain; but they occur at a much earlier period and to a much more intense degree with tumors situated at the base of the brain. Under these conditions the direct pressure upon the optic chiasm and the greater increase of intra-cranial fluid at the base may account for the earlier appearance of the symptoms. The optic neuritis is generally double, though it may be more distinct for a time in one eye than in another.

Sudden variations in the pulse-rate, and in rapidity of respiration have been observed; Cheyne-Stokes respiration I have seen in children suffering from cerebellar tumors long before the terminal stage had been reached.

The presence of tumor can often be suspected from the occurrence of these general symptoms. The signal or localizing symptoms are dependent upon the special functions of the areas invaded by the neoplasm. These localizing symptoms are the best illustrations in man of cerebral functions, as they have been demonstrated by experiments upon animals. The matter is not quite as simple, however, as it would seem, for a neoplasm causes both direct and indirect symptoms. The direct symptoms are those caused

by the actual loss of function due to displacement of tissue by the neoplasm. The indirect symptoms are caused either by the distant effects of the pressure by the tumor upon neighboring parts, or they may result from the secondary increase of intra-cranial fluid. At all events, the symptoms first produced are the most important ones, and will be of the greatest value in determining the exact site of the lesion.

The indirect symptoms are perhaps more frequent in the cases of cerebellar tumors than in any other. Thus in this form we may have the general symptoms of intra-cranial neoplasm, together with reeling and distinct occipital headaches. Associated with these we often find a paralysis of the rectus externus supplied by the sixth nerve. Spinal symptoms may occur with cerebellar and other intra-cranial growths. Batten and Collier, Anton, Pick, and a number of German writers have referred to the frequent involvement of spinal cord structures (the posterior roots and posterior columns), evidently due to the increased pressure of the cerebro-spinal fluid.

MACEWEN'S SYMPTOM.—"The elicitation of a differential cranial percussion-note as an aid to cerebral diagnosis in certain gross changes of the intra-cranial contents, especially in children," has been practised by Macewen for many years and described in his monograph on "Infective Diseases of the Brain and Spinal Cord." For the present, the symptom is most valuable in the case of abscesses and of over-distended lateral ventricles. In cases of tumor near the surface of the brain the percussion-note will prove of service. I have noted a dulness in several cases of tumor of the brain (one case in the adult and two in children); if the lateral ventricles are over-distended the resonance is greatly increased. But distention of the ventricles may be secondary to tumor, and in this way increased resonance of the percussion-note may be significant. The percussion-note of normal crania seems to vary much; it will be best, therefore, to use this symptom very cautiously. The note can be elicited best by tapping the skull lightly with the finger or percussion-hammer, and receiving the note through a stethoscope placed upon the pterion, or upon the middle of the forehead.

TUMORS OF THE CORTEX.—It is not an easy matter to distinguish between the purely cortical tumors and those situated in the subjacent white matter. Some claim that it is impossible to differentiate between cortical and subcortical tumors, but this seems to me to be rather overstating the facts. While the localizing symptoms may be exactly the same in both these classes of tumors, the order of development of the symptoms will be different, and may give some indication of the exact site of the tumor. In subcortical tumors the march of the symptoms is generally such that the approach of the lesion toward the cortex is marked by a special series of symptoms. Taking, for example, the tumors in the motor area, those in or near the gray matter, however small, are apt to give rise to occa-

sional convulsive seizures from the very start; whereas, in the tumors which begin in the subjacent white matter, the paralytic symptoms may appear at a very early day and may exist for some time before any symptoms of cortical irritation arise. Thus, in a case under my observation, the patient had gradually developed paralysis of the arm, of the face, and slight paresis of the leg on one side of the body. He had indistinct headaches and optic neuritis, and yet, during the first three months after the onset of these symptoms, not a single convulsive twitching occurred. Since that period epileptoid seizures have set in, and the occurrence has proved that the tumor has finally invaded the cortical region. The invasion of the cortex, moreover, is more likely to lead to repeated convulsive seizures and to increased headaches from the more direct involvement of the meningeal coverings; the nearer the surface and the nearer the meninges the more intense these headaches become.* Certain classes of tumors, moreover, are more apt to begin in the cortical tissue than in the subcortical white substance; thus tubercles, gliomata, and gummata, above all, are almost certain to begin near the surface, while sarcomata and cysts are as often subcortical as cortical in origin.

TUMORS OF THE FRONTAL LOBE.—Since the frontal lobe has no distinct and special function, tumors in this region are often developed without localizing symptoms. If the tumor extends downward and involves the olfactory bulb the sense of smell may be diminished or lost; but changes in character and intelligence, "punning," irritability or stupidity, are considered to be relatively frequent in diseases of the frontal lobe, and this is entirely in support of the conditions which Goltz first discovered as a change in the behavior of dogs whose frontal lobes were excised or destroyed. More recently Torje has recorded a case in which there was marked disturbance of salivation; he suggests that the drooling in idiots may be due to defective frontal development. Bruns has mentioned the occurrence of cerebellar ataxia in these cases. Frontal tumor may also be diagnosticated if the general symptoms were present for a long period of time, and then symptoms pointing to the third frontal gyrus or the motor areas appear as evidence of the gradual encroachment of the new-growth upon these areas.

TUMORS OF THE THIRD FRONTAL CONVOLUTION.—The third frontal convolution is the seat of motor speech function, and any destructive lesion in or near this part of the brain will cause typical motor aphasia. Agraphia may be associated with motor aphasia. The cases in which motor aphasia, or agraphia, or both, have signalized the invasion of a tumor are comparatively rare, particularly in children; for if the invasion of the tumor be a gradual one the right hemisphere may, in right-handed persons, gradually assume the functions of speech, which up to that time have been inherent in the left hemisphere. In addition to this, in young children the differentiation between the left and the right hemispheres is not nearly as complete as in later years.

TUMORS OF THE MOTOR AREA.—These can be recognized more readily than those in any other part of the cortex. In the earlier stages of the dis-

* If there is excessive intra-cranial pressure, headaches may be intense, even in tumors distant from the surface.

ease symptoms of cortical irritation are present, and convulsive twitchings of the paretic or paralyzed part will indicate the probable site of the tumor. Thus, if a patient with slight sensory and motor disturbances in the upper extremity should experience occasional convulsive twitchings of the arm, of the forearm, or even of the thumb alone, the character and extent of these twitchings would point to the part of the brain first and chiefly involved. According to the course and development of the convulsive seizures the direction in which the growth extends can be judged. Vasomotor symptoms and changes in pulse-rate are uncommon.

TUMORS OF THE PARIETAL LOBE.—Tumors of the parietal lobe may cause no distinct localizing symptoms, but the records of Mills and Spiller prove conclusively that lesions in these parts are apt to be followed by sensory changes (loss of muscular sense, astereognosis, etc.). These facts of human pathology are in keeping with the results of Munk's experiments on dogs, and those of Schafer and Horsley on monkeys. These sensory symptoms, however, are not so constant that their absence would militate against the diagnosis of tumor in this region. If a tumor in the parietal lobe affects the subjacent white matter it may result in hemianopsia, for the white tract of Gratiolet passing from the internal capsule to the occipital lobe lies under this portion of the cortex. Ferrier's claim, that the centre of vision lies in the angular gyrus, is due to the involvement by disease in this region of the optic radiations which we have mentioned. By extension to the inferior parietal lobule the tumor may give rise to that peculiar disturbance of speech known as word-blindness. The patient cannot read and write at will, though he may be able to write upon dictation, or to copy written signs. In children, excepting those of a more advanced age, cases will rarely arise in which word-blindness or word-deafness are important symptoms. According to Wernicke the conjugate movements of the eyes are governed by a centre in the inferior parietal lobule. If there is distinct impairment of this one function, disease in this region may therefore be suspected.

TUMORS OF THE OCCIPITAL LOBE.—Tumors in the occipital lobe are recognized by the peculiar disturbances of vision. As each occipital lobe is connected with one-half of each eye, tumor in this region is signalized by homonymous hemianopsia without any other special symptoms; convulsions may occur, and not necessarily on account of the direct extension of the lesion into the motor areas, but because, as has been shown by Binswanger, disease in any part of the cortex may cause epileptic disturbances. The researches of Nothnagel and Seguin, which have been corroborated by other authors, points with a great degree of certainty to the cuneus as the actual centre of vision. Visual hallucinations and word-blindness have been recorded by a number of reputable writers. According to Bruns, optic neuritis is relatively infrequent.

TUMORS OF THE TEMPORO-SPHENOIDAL LOBE.—The temporo-sphenoidal lobe contains the centres for hearing and for sensory speech; it is, therefore, natural to expect that in the case of tumor in these auditory regions hearing will be impaired, not abolished, on the side opposite the lesion, and sensory aphasia will be present. The patient will be able to speak spon-

taneously and correctly, but will not be able to understand what is being said to him, and will, of course, not be able to repeat spoken language. The differential diagnosis between tumor and abscess should be established carefully. Deep-seated tumors may cause hemianæsthesia, hemiplegia, and hemianopsia.

TUMORS OF THE BASAL GANGLIA.—Tumors of the basal ganglia and the adjacent parts are not rare. The symptoms which they produce are, as far as we know, entirely due to direct or indirect involvement of the internal capsule. As this capsule contains the entire motor tract for the opposite side of the body, the sensory fibres, the fibres of special sense, the speech-tracts, we can infer the multiplicity of symptoms which may result from such a lesion. The position of these fibres in the internal capsule is quite well established, and in the case of small tumors, the order of involvement of the different functions may give some clew as to the direction in which the tumor is extending. The tumor may impinge, too, upon the lateral ventricles, and thus cause considerable disturbance. Some of the symptoms may be secondary to the increase of hydrocephalic fluid. On account of the compact structure of the inner part of the brain, even small tumors will cause considerable distortion of the parts, and in addition to all the other symptoms presented as the result of the involvement of the internal capsule, we may have symptoms resulting indirectly from pressure upon the cranial nerves, which come off from the brain axis below this region. The differential diagnosis between tumors of the ganglia and tumors of the cortex will be based in part upon the absence of convulsive seizures in the case of ganglionic tumors, and the more frequent presence of choreic or athetoid movements with cortical disease.

TUMORS OF THE CRUS CEREBRI.—A neoplasm in this region can be recognized very early by the association of oculo-motor symptoms with paralysis of motion and sensation of the opposite half of the body. The eye will exhibit ptosis, paralysis of all the external muscles except the rectus externus and the superior oblique, and complete paralysis of the sphincter iridis, and the ciliary muscle.

Both peduncles are so close to one another that a tumor occurring in one may actually invade the opposite side, or at least press against the peduncle of the other half of the brain. For this reason it is not uncommon to find paralysis of both halves of the body, or possibly double ptosis and double oculo-motor symptoms in tumors in this region. In the majority of cases, however, the symptoms are strictly unilateral for a long period, and become bilateral later on.

TUMORS OF THE CORPORA QUADRIGEMINA.—These tumors should in reality be considered with tumors of the peduncles, for these structures are so closely related to one another, anatomically and physiologically, that the occurrence of neoplasm in the one part or the other will produce a common series of symptoms. It is only the occurrence of oculo-motor paralysis with opposite hemipleglia that suggests the region of the peduncles, while a few additional symptoms refer distinctly to the corpora quadrigemina. These additional symptoms are due to the relation which the corpora quadrigemina

bears to visual functions, and to the connection between the former and the cerebellum. Loss of pupillary reflexes, nystagmus, vertigo, and a condition resembling cerebellar ataxia, point to the region of the corpora quadrigemina as the special seat of the lesion. I had occasion a few years ago to publish an interesting case of this description, with the result of the post-mortem examination (Fig. 120). Tumors of the pineal gland will cause an entirely similar series of symptoms. Marked vasomotor disturbances have been recorded by Collins and others with neoplasm in this region.

TUMORS OF THE PONS.—Tumors of the pons and medulla, like other lesions in these parts, give rise to a great multiplicity of symptoms. The pyramidal tracts are closely approximated to one another near the median line, so that, though the symptoms may be unilateral, they are often bilateral. In addition, therefore, to the hemiplegia, or double hemiplegia, we may have other symptoms pointing to an involvement of the various cranial nerves. A neoplasm in the upper half of the pons may encroach more upon the peduncles than upon the structures of the pons itself, resulting in a hemiplegia of one side of the body with an involvement of the third and fifth nerves of the opposite side. If the tumor is in the lower half of the pons, the fifth, sixth, seventh, and eighth nerves will be more or less involved, and the symptoms resulting from this affection will be paralysis of the rectus externus, paralysis of all the branches of the seventh nerve in one half of the face, and a loss of hearing in one ear. All these cranial nerve symptoms will be on the side of the lesion opposite the hemiplegia. If the sixth nerve nucleus is affected there will be, in addition, distinct paralysis of one rectus externus muscle, and paralysis of the conjugated movements of the eyes toward the side of the lesion, for this nucleus is connected with the third nerve nucleus of the opposite side and governs the outward movement of each eye. In spite of this conjugate paralysis each internal rectus if examined separately may exhibit normal movements. If the lesion is near the surface of the pons, and away from the nucleus, it will involve the root of the sixth nerve, and will cause paralysis of the rectus externus muscle of one side, but will not affect the conjugate

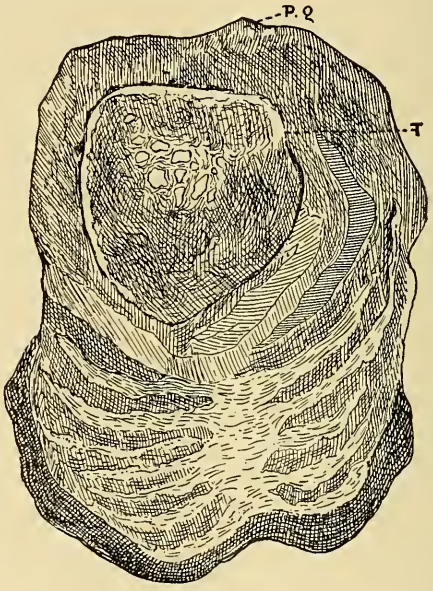


FIG. 120.—Section passing through the Posterior Quadrigeminal Bodies. T, tumor (solitary tubercle); P. Q, posterior quadrigeminal body.

movements of the opposite side. If the patient is directed to look toward the side of the paralyzed rectus or toward the side of the tumor the opposite eye will move promptly, the affected eye remaining fixed. This differentiation between the isolated paralysis of the rectus externus and paralysis of the conjugated movements is the most valuable, and perhaps the only differential point of diagnosis that helps at times to distinguish between a tumor near the surface and one within the substance of the pons.

The difficulties of diagnosis of pons lesions are increased by the exceeding variability of the symptoms, and by the fact that some of the nerves in the pons escape disease, whereas others may be intensely affected.

In the case of a tumor in the medulla oblongata the symptoms are very similar to those met with in bulbar palsy. The symptoms will indicate disease of the glosso-pharyngeal, of the vagus, of the spinal accessory, and of the hypoglossal nerves, together with a unilateral or bilateral paralysis of the arms or legs; the facial nerve will not be involved. Difficulties in deglutition, in respiratory and cardiac movements; paralysis or spasm of the sterno-cleido mastoid, and of the trapezius; paralysis and atrophy of the tongue, together with vomiting, with glycosuria or polyuria—all these signs will suggest a lesion in the medulla. Tumors in this region, and particularly gummata, are not rare. Difficulties in diagnosis often arise on account of the bilateral character of all the symptoms, but it should be remembered that the two halves of the brain at this level are scarcely separated from one another, and that it is natural for all but vascular lesions (and even these may do so) to produce bilateral symptoms. In the pons and medulla the sensory tracts also lie very closely to the motor, and for this reason the symptoms may be still further complicated by the occurrence of partial, or complete, or double hemianæsthesia. As the cerebellum is also in close juxtaposition to the medulla and the pons, symptoms of cerebellar involvement may be present in the cases of neoplasms, both in the pons and in the medulla. Optic neuritis is developed at a very early day, and occipital headaches are particularly severe.

TUMORS OF THE CEREBELLUM.—Tumors of the cerebellum are the most frequent. The symptoms are occipital or frontal headache, projectile vomiting, rigidity of the neck, and (at a relatively early period) a peculiar staggering or reeling gait (cerebellar titubation). Double optic neuritis has been found in eighty-nine per cent. of these cases. Inco-ordination is frequent. Asthenia or muscular weakness is a real cerebellar symptom (Mills). Asynergia or hemiasynergia (Babinski)—asynchronous movement of the thigh and leg in extending the limb—is of questionable value. Vertigo is also more frequent and more severe in cerebellar tumors than in many others. In my own rather extensive experience I have found that the patient is apt to fall *toward* the side on which the tumor is located. Mills believes that destructive and irritative lesions of the vermis are responsible for a tendency to fall forward or backward. A cerebellar tumor may be of very small size and yet sufficiently large to compress the sixth, seventh, and eighth nerves, and in its further growth may produce involvement of the bulbar nerves as well, just as tumor of the bulb may also produce cerebellar symptoms. The affection of

the sixth nerve, causing paralysis of the rectus externus, is extremely common in cerebellar tumors, and in some cases the diagnosis of cerebellar tumor may be corroborated by the early involvement of the seventh and eighth nerves (Fig. 121).

Of late years, Monakow, Sternberg, Oppenheim, Fraenkel, and Hunt have called attention to slowly growing tumors (neuromata and neurofibromata) in the cerebello-pontile angle, involving one or more cranial nerves, especially both auditory nerves. Cerebellar symptoms are preceded, often for many years, by those pointing to involvement of the auditory, the trigeminal, the

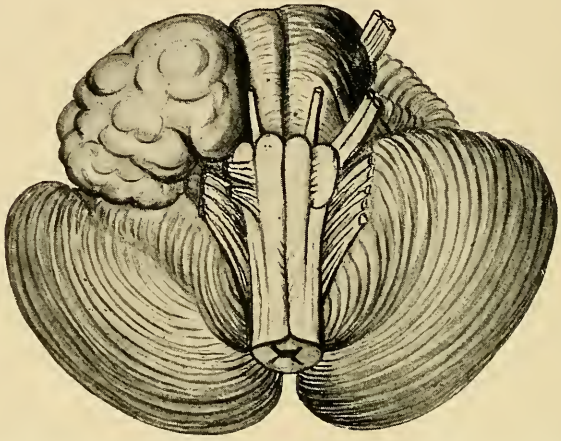


FIG. 121.—Tumor (Sarcoma) of the Cerebello-Pontile Angle. Early appearance of facial and auditory nerve symptoms.

vagus, of one or both sides. I have under observation at the present time a young girl who, for years past, has suffered from central deafness, ataxia of the right upper extremity, dizziness, and who now is also completely blind from double optic neuritis.

The general manifestations of cerebellar tumors do not vary much from tumors in other parts of the brain; but if the growth encroaches upon the pyramidal tracts in the pons and medulla unilateral or bilateral paralysis will follow. The reflexes are sometimes exaggerated, as in all other cerebral diseases, but in not a few cases the knee-jerks are either diminished or absent.

DIFFERENTIAL DIAGNOSIS.—The diagnosis between tumor and abscess will depend upon the frequent presence of fever, and upon the very slow invasion and slow development of all the symptoms in the latter condition, also upon the occasional absence of optic neuritis in spite of the presence of symptoms pointing to increased intra-cranial

pressure; but the differential diagnosis will depend first and foremost upon the proof of the presence of such conditions which give rise to abscess, or the absence of such conditions in the case of tumor. For other symptoms characteristic of abscess rather than of tumor, the student is referred to the next chapter.

The ordinary forms of tubercular meningitis are so distinct that they cannot be readily confounded with tumor of the brain, but the difficulty lies in the fact that some cases of tubercular affection of the brain take an exceedingly slow course, and that some forms of tumor prove rapidly fatal. Furthermore, tumor in the form of solitary tubercle and meningitis are occasionally associated with one another, as was proved in a case of mine of solitary tubercle of the corpora quadrigemina. In meningitis, whether of a tubercular character or not, the rapid involvement of a number of cranial nerves, without an increase in all the other symptoms, will argue in favor of this disease rather than of tumor. This is true also of the specific meningitis as well as of the tubercular form.*

Histological examination of the cerebro-spinal fluid, obtained by lumbar puncture, may assist in the diagnosis. Beware of lumbar puncture in cerebellar disease! The X-rays may occasionally be of service in locating a new growth (Mills, myself and others).

PATHOLOGY.—The most important intra-cranial growth is the solitary tubercle. It occurs both in children who exhibit distinct tubercular tendencies, and also in those who neither give a family history of tuberculosis nor exhibit any cachectic symptoms.

In size the solitary tubercle may vary from that of a pea or cherry to one as large as that depicted in Fig. 122. In this brain it will be seen that a solitary tubercle had occupied the greater part of one hemisphere, and had considerably distorted the entire brain axis. Very often the tumor starts in the vicinity of the meninges, or at least near the cortical blood-vessels. The blood-vessels unquestionably play an important part, for in cases of tuber-

* Oppenheim has reported several interesting cases of children presenting all the symptoms of a tumor of the Rolandic area, and ending in recovery. He thinks these may have been cases of meningo-encephalitis tuberculosa. Nonne has also described cases in adults and in youthful individuals supposed to be suffering from tumors of the brain; all symptoms had receded. Hydrocephalus, serous meningitis, and specific lesions may cause difficulties in diagnosis.

cular meningitis of the base smaller tubercles of a pin-head size may be seen scattered along the vessels on the outer surface of the brain. A solitary tubercle is composed of a thick stroma with giant cells and a considerable amount of hard fibrous tissue. There is a more or less concentric arrangement of the parts, and the central portion of the tumor, in many cases, undergoes caseous degeneration at an early day. As in the cord, so in the brain, the tissue surrounding a tubercular growth may break down and become an almost diffluent mass. A tubercular encephalitis, or meningo-encephalitis, may, therefore, be superadded upon a solitary tubercle. In just as many cases, however, the solitary tubercle behaves as every other cerebral tumor does. It is, as a rule, sharply differentiated from the surrounding tissue, and

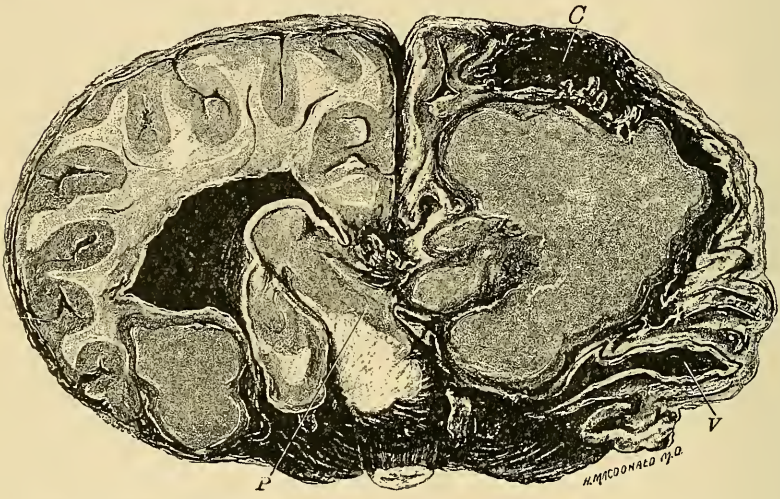


FIG. 122.—Vertical Section through Cyst, *C*, and Both Tumors, showing Distortion of Brain Axis and Displacement of Left Ventricle. *P*, pons; *V*, the left ventricle.

the exact character of it can be made out only on histological examination. The very size of the tumor, as in the brain depicted in Fig. 122, is suspicious of solitary tubercle, for the smallest as well as the largest tumors occurring in the brain are of this character. In the histological examination of the tumor search should be made for the tubercle bacilli, and their presence will at once prove or disprove the supposed character of the tumor.

Glioma and sarcoma, or a combination of both, are the next most frequent forms of neoplasm occurring in the brains of children. The glioma possesses the characteristics of neuroglia tissue. Its growth is extremely slow, and for this reason it may be suspected in cases in which all the symptoms point to a period of growth lasting for one or two

years, or even a longer period of time. In connection with glioma, as in other forms of tumor, there is, as a rule, a considerable increase in the blood-supply to surrounding parts. An increase of cerebro-spinal fluid is a constant accompaniment, and it must be due to variations in the amount of these fluids that improvement sometimes follows upon treatment by iodides and mercurials.

A patient of mine had been suffering for a long time from symptoms pointing to a tumor in the parietal region; he had also had an apoplectic attack some years previously. Taking these facts into consideration I concluded that the tumor was probably of a specific character, and placed him upon a very rigid iodide and mercurial treatment. The improvement was most marked, the headaches ceased, the convulsive seizures disappeared altogether, and the power in the right hand returned. I was convinced of the specific character of the tumor. The patient died, however, in a relapse, accompanied by an apoplectic seizure, and to my great surprise, on post-mortem examination, the tumor, which was examined by Dr. Van Gieson, proved to be a glioma and not a gumma. It is of the utmost importance, therefore, not to attach too great significance to any improvement occurring in these diseases after specific treatment.

Gliomata occur more frequently in the white than in the gray matter of the brain, but may invade the latter though starting from the white substance. The pons shares with the hemispheres the distinction of being the most frequent seat of glioma, the dense character of the tissue in the pons evidently yielding favorable conditions for the development of this special form of neoplasm.

Sarcoma, pure and simple, or in connection with glioma, is somewhat less frequent than glioma alone. Its growth is more rapid than that of simple glioma, and it is not as distinctly separated from the healthy tissue as glioma generally is; the tumor and infiltration as a rule extend for some distance into the neighboring tissue. By this invasion and by actual compression much of the tissue in the vicinity of a sarcoma is readily broken down. Round cells and spindle-shaped cells occur together with the fibrous mass, and according to the predominating character of the cells the sarcomata are divided into round-celled and spindle-celled varieties. Myxo-sarcoma is not unknown in childhood, and a few cases have been reported of that much rarer form known as melano-sarcoma. The sarcomata are invariably secondary to similar diseases in other organs. Sarcoma may therefore be suspected with malignant disease in other parts of the body, and particularly if the tumor be one of rapid growth, and if the symptoms point to an intensely destructive process.

Cysts are extremely frequent in children, and are often found in brains not suspected of any gross disease. Such cysts are generally the remnants of an acute process early in life, an early hemorhage, or a softening due to

embolism or thrombosis. These cysts fill up with fluid, and cause in the majority of cases no symptoms such as we assign to tumor; but if from any cause such an old cyst is lighted up and its contents suddenly increase, the symptoms may resemble those of cerebral neoplasm. It is well to be prepared for the occurrence of such cysts, and to know that they are a favorite starting-point for other tumors. It was on the strength of such familiarity with the cerebral conditions of childhood that I was able to make the diagnosis of a tumor growing from the cyst-walls in the case referred to on page 569.

Cystic tumors, in the true sense of the word, are those which are due to an invasion of echinococcus. In this country it is extremely rare to find a tumor of this description, but in Europe and in Eastern countries they are far more frequent. The cysticercus cysts occur in children as well as in adults, are multiple, and may give rise to a confusion of symptoms.

The only other form of neoplasm which deserves special consideration is that due to syphilitic disease Gummata are unquestionably rare in children, but they may of course occur in cases of hereditary syphilis. We should look for them in connection with specific disease of the brain and its coverings. Repeated attacks and relapses would suggest the syphilitic character of the disease.

Aneurism is also so rare in childhood that it would hardly need to be considered. But Osler reported some years ago an interesting case of aneurism in a child six years of age. The symptoms are very much like those caused by ordinary neoplasm, but the course of the disease is much more chronic than in the case of other new-growths. The diagnosis of aneurism could be reached only by exclusion, by the slow course of the disease, and possibly by irregularities of the pulsations of the two carotid arteries. The anterior and posterior cerebral arteries are the vessels favored by aneurisms of any considerable size.

In concluding these remarks upon the pathology of tumors, it is well to remind the student once more that the symptoms produced by any tumor are in part due to the tumor itself, to the character of the invasion, and to the region of the brain affected, and in part to the pressure upon the neighboring tissues, and upon distant parts of the brain structure. The increase of intra-cranial fluid plays a most important part in the causation of some of the symptoms, notably of headaches and of optic neuritis.

TREATMENT.—In spite of many advances in recent years, the medical or surgical treatment of brain tumors has been

disappointing. According to Starr's statistics only 7.5 per cent. of the cases are operable. Ferrier and Oppenheim give a similar percentage. Bramwell thought many years ago five out of twenty-two cerebral tumors could have been removed. In children the prognosis is still gloomier. For this there is a double reason; first, tubercle is very much more frequent in children than in adults; second, if the conditions for the removal of the tumor are favorable, the child stands a poorer chance of surviving the operation than the adult does. Before proceeding to the surgical treatment of the case of brain tumor, which is, after all, the only thoroughly rational course, the attempt should be made to influence the growth, and to promote the absorption of the new-growths by various drugs. The most powerful drugs are the iodides and the mercurials. Gummata are very rare in children, and this treatment would therefore be of little use if it were given for its anti-syphilitic effect. But in every case of tumor there is considerable exudation, considerable increase of cerebro-spinal fluid, and both the iodides and the mercurials seem to have some effect upon the absorption of these products of inflammatory reaction in connection with tumor. By promoting such absorption of adventitious products the intense localized headaches and persistent vomiting, and other general cerebral symptoms frequently present, are modified. Another reason for the employment of specific remedies is the belief that if they will not avail any in the actual cure of the trouble, they will at least help to relieve some of the most distressing symptoms that occur in connection with cerebral neoplasm.*

If any decided effect is to be hoped for, the iodides, as well as the mercurials, should be given in effective doses; the iodides in doses varying from five to twenty-five grains three times a day; the mercury, in the form of inunctions, from fifteen to thirty and forty-five grains twice a day, according to the age of the child. All other medicinal treatment will be of no avail.

The headaches will be relieved best by a combination of phenacetine with codeine (one-fourth to one-third of a grain), or with small doses of morphia (one-sixteenth to one-eighth of a grain), according to the age of the child. Morphia alone I am not in favor of giving; and if the pains are so extreme

* Lumbar puncture might be tried in inoperable cases for the relief of pressure. Trephining for the relief of pressure is justifiable and beneficial.

that the child becomes restless and sleepless in consequence of them, I should prefer moderate doses of chloral hydrate, from five to ten grains, according to the age of the child, given either per mouth or per rectum. If the tumor causes frequent convulsions the bromides might be added to the chloral, but it is best to avoid them in all cases in which epileptic seizures do not occur, for the bromides have no effect upon the intense headaches, unless given in very large doses, and have a disagreeable effect, interfering with the digestion and nutrition of the child. The ordinary hypnotics, such as sulfonal, trional, veronal, and the like, can be employed in moderate doses in those cases in which there is a general restlessness without pain.

In the first edition of this book the author took a distinctly pessimistic view of the surgical treatment of brain tumors. Within the last few years the brilliant work of Horsley, of Kocher, of Keen, and the recent enthusiastic and ingenious writings of Frazier and of Cushing have given promise of far greater successes in this special field. Even the cerebellum has been made accessible. Sänger and Cushing are right in urging the great value of a palliative craniectomy as a means of "decompression" for the relief of headaches, and for the preservation of the eyesight. Within the last year this palliative operation has been performed for me by Gerster or Lilienthal in four cases of cerebellar tumor occurring in children. The difficulties of surgical interference in children are, in part, similar to those met with in the adult, and in part peculiar to children only. Among the first we may mention the uncertainty which still prevails in every case regarding the exact size of a tumor, particularly if the tumor begins in one of the active areas of the brain and spreads to a so-called silent area. On this account alone, however, no one would be justified in refraining from an operation; this uncertainty constitutes one of the attending risks.

Among the conditions peculiar to children I would mention first of all the fact that children, according to my experience, which has been quite extensive, tolerate operations upon the brain very poorly indeed. The cerebral shock, and, above all, the considerable cerebral hemorrhage, lessen the chances of recovery very much.* If we consider, moreover, that a very large number of the cerebral tumors in children are of a tubercular nature, that multiple

* The student is referred to the interesting articles of Frazier (N. Y. Med. Journal, February 11 and 18, 1905) and of Harvey Cushing (The Johns Hopkins Hospital Bulletin for March, 1905).

tumors are not infrequent, that the tumors occur with great frequency in the cerebellum and on the ventral surface of the brain, we are forced to the conclusion that there are but few cases of intra-cranial tumor in children which seem favorable for operation, and that of these few it must be a rare fortune indeed to have a single one recover completely. It is the duty of every careful physician to weigh the evidence for and against an operation in every case, and if in a child of good physical strength a non-tubercular tumor is suspected in a region accessible to the surgeon's knife, the attempt at removal of such a tumor is justifiable if all other remedial measures have been given a previous trial. Whether a tubercular tumor should be removed or not is a question that cannot be decided off-hand. The answer may possibly be given some day by the results of operation in a case in which the character of the tumor was not suspected until it had been successfully or unsuccessfully removed.*

* v. Beck has reported a case of tubercle of the brain successfully removed by Czerny. The author reports improvement after the operation. Some years ago Dana exhibited a boy of about 12 years old who had done well for more than a year after the partial removal of a sarcoma.

CHAPTER XXVII.

ABSCESS OF THE BRAIN.

AN abscess may be situated either on the surface of the hemispheres, or within the substance of the brain. The latter location is by far the more frequent. Multiple abscesses sometimes occur. Bergmann found one hundred or more abscesses in a single case following gangrene of the leg. In one case the author observed at least six abscesses, which were successfully incised; but a single abscess is more common. If the pus is on the surface of the brain the membranes, which are generally thickened as the result of the inflammatory process, may constitute one wall of the abscess.

Abscess of the brain may occur at any period of life. Of the cases which Gowers has tabulated fully one-third occurred before the age of nineteen. The male sex is more disposed to this disease than females, in a ratio varying from two to one to five to one, according to the initial cause. The most frequent cause of abscess of the brain is preceding ear disease. Next in frequency is external injury to the skull. Gull and Sutton, in their article on abscess of the brain, in Reynolds's "System of Medicine," state that 102 cases of abscess were due to ear disease, and 57 to injury. Koerner, in his exhaustive monograph on the otitic diseases of the brain, quotes Pitt's statistics, who records 56 cases of brain abscess among 9,000 autopsies. Of these 56 abscesses 18 (very nearly one-third) were due to disease of the ear and of the petrous bone, while 8 (one-seventh of the entire number) were due to disease of other cranial bones. The relative number of other cerebral conditions complicating ear disease, as given by Koerner, is as follows: Phlebitis and thrombosis of the sinuses occurred in 44 cases among 9,000 autopsies, and of these 44 fully one-half were due to disease of the bony parts of the ear. A simple otitic meningitis as the result of purulent ear disease is relatively rare. Taking all cases of cerebral disease complicating otitis into consideration Koerner states that of 246 cases which he has been able to collect, 44 occurred before the age of ten, and 73 between the ages of eleven and twenty years. Thus it will be seen that fully one-third of all abscesses occurred during the period of childhood and early youth. Koerner

states, basing his assertions upon Prussian statistics, that 5.15 per cent. of all deaths between the ages of ten and twenty were due to cerebral disease complicating otitis. These statistics will suffice to indicate the great interest that must attach to suppurative disease in early life, and particularly to abscess of the brain following upon disease of the ear. Other probable causes of abscess must not be neglected. Disease of the nose, empyema of the chest, and accidental injury to the skull, whether or not it has given rise to a tangible bone lesion, may be the starting-point of brain abscess.

Gowers states that fifteen per cent. of all the cases of abscess of the brain were due to what he calls distant influences. Among these any condition giving rise to a general infection may be included, and it is more than probable that even among this fifteen per cent. of cases some may have been traumatic, although the exact character of the traumatism could not be determined.

In abscess of the brain following upon chronic otitic disease the ear trouble may have existed for several years. I have known such ear disease to continue for ten years before the symptoms of abscess were developed. As long as there is a free discharge of pus outward, the danger of cerebral disease is comparatively slight; but if the discharge is checked, the pus may burrow its way through the thin plate of bone separating the middle ear and the mastoid cells from the cranial cavity. The small veins which pass through the ear structures into the cerebral sinuses (superior petrosal and the lateral sinus) provide easy means of access for the pus from the inner ear into the cranial cavity. From the mastoid cells the pus passes most easily into the lateral sinus. In connection with ear disease the abscess generally forms either in the temporo-sphenoidal lobes or in the hemispheres of the cerebellum. Usually it affects these regions, on the same side of the brain. In rare instances the chief formation of pus is on the side opposite to the diseased ear.

The abscess formed in connection with ear disease is more often underneath the gray matter of the cortex than upon the outer surface of the hemispheres, and not infrequently normal tissue separates the abscess from the carious bone and the meninges. Such being the case, it is evident that the abscess must have been formed within the brain and gradually extended outward. In a few instances a direct path of connection between the suppuration in the ear

and the abscess cavity has been demonstrated, but in the vast majority of cases no such connection has existed. The problem, therefore, is to decide in what manner the pus-corpuscles could have been carried from the ear to the interior of the brain. No entirely satisfactory solution has as yet been given, but the frequent interference with the circulation in the sinuses undoubtedly plays an important part. If we take into account the relative position of the temporo-sphenoidal lobes and the structures of the ear it will be seen that the superior petrosal sinus receives blood from both these parts, and, on the other hand, the lateral sinus receives blood from the cerebellum and the mastoid cells. It is due to these venous connections that abscess of the cerebellum is met with more frequently in connection with mastoid disease, and abscess in the temporo-sphenoidal lobes more frequently with disease of the middle ear. Such abscesses have been reported by Lucae, Koerner, Bruns, Macewen, Gruening, Bacon, Dench, and many aural surgeons.

Abscess of the brain in connection with diseases of the nose must be explained in much the same way (Dreyfuss). The nasal disease is sometimes limited to the mucous membrane without any involvement of the bone, yet abscesses form just the same. Under such conditions the toxic substance must be carried along the blood-vessels or the lymphatics. If the nasal bones are the seat of suppuration, as in syphilis, the abscess that forms may be in direct connection with the nasal cavity, and the pus from the brain may be freely discharged through the nose.

The breaking down of tubercular growths may occasionally be the cause of abscess, but these cases have less practical interest, for the general tubercular character of the disease is the significant feature, and the abscess, as a rule, a mere incident in the course of a protracted illness.

Purulent disease in any other part of the body may also be the cause of abscess of the brain through the agency of a septic embolus. Gowers quotes Boettcher's report of a case in which a cerebral abscess secondary to suppuration in the lung was found to contain lung pigment. There is a remarkable connection between brain abscess and every form of purulent disease within the thoracic cavity. Thus we have cases of brain abscess after pneumonia in which the exudate has not been perfectly resolved and has undergone suppuration. Pus cavities occur after purulent bronchitis and empyema; recently Moschowitz operated upon a young boy in whom the author had helped to locate an abscess in the Rolandic area following empyema.

As a rare cause of abscess, but one that is worth mentioning, we may note its occurrence after thrush in two cases reported by Zenker. One patient was an infant. In these cases the brain was studded with small abscesses con-

taining the fungus of thrush (oïdium albicans); the fungus had evidently travelled by way of the blood-vessels, since it had actually been demonstrated in their interior.

SYMPTOMATOLOGY.—Few conditions are more difficult to recognize during life than abscesses of the brain. It is safe to say that they constitute a large proportion of the surprises and disappointments of the post-mortem table. They are found when least expected, and when confidently looked for are often wanting. The uncertainty in the diagnosis is due to the very insidious development of the process and to the fact that when once encapsulated a pus cavity gives rise to very few symptoms.

When fully developed an abscess practically constitutes a foreign body in the brain; we would therefore expect such symptoms as are commonly due to tumors within the brain or cranial cavity, but in this we are apt to be disappointed, for the brain appears to accommodate itself to the slowly increasing abscess and does not respond to this growth as distinctly as it does to the invasion of a solid neoplasm. The abscesses do not, moreover, occur as frequently as tumors do in those parts of the brain which give rise to signal symptoms (the motor area and the occipital lobes). Abscesses are more common in the frontal lobes, in the temporo-sphenoidal lobes, and in the cerebellum. In acute cases, favorable for diagnosis, the first symptoms are those with which we are familiar in connection with meningeal disturbances, such as local painfulness of the scalp, nausea, vomiting, vertigo, and fever. The last, if irregular and if attended by rigors, is the one symptom which suggests abscess much more forcibly than any other. If the cavity is in the vicinity of the motor areas and is not too rapidly destructive, localized convulsions may occur. Abscess in this region, which is supplied by the middle cerebral artery, is very likely to be due to septic embolism; hence the formation of pus in this region after lung disease and after septic endocarditis. Paralysis may be present in some of these cases if the motor area is involved, but if the abscess is elsewhere in the brain paralytic symptoms will not constitute a prominent feature of the disease until the terminal stage has been reached.

To this category of symptoms, delirium, convulsions, irregular or high temperatures may be added, which lead to a rapidly fatal issue.

Chronic abscess may exist for a number of years and may give rise to very few symptoms; in one case of chronic abscess, in a boy of eight years, no symptoms were present as long as the ear freely discharged. As soon as this ceased, epileptiform convulsions, preceded by sensory auræ (disagreeable odor and a hissing noise) would occur. This alternation was kept up for at least two years, when the boy died of a general purulent meningitis from bursting of the abscess into the temporo-sphenoidal lobe. The so-called latent state of the abscess is characterized, as a rule, by occasional spells of nausea, by occasional vomiting, and by intermittent febrile attacks for which no proper explanation can be found at the time. Persistent headaches, if associated with ear disease or with other conditions that give rise to abscess, point to the existence of this condition. The headache is generally more or less circumscribed, and sometimes varies according to the position of the head; but this mere fact of change of position as affecting the headache does not necessarily point, as one would suppose, to abscess, for I have known the same condition to accompany solid tumor of the brain. The pain, as a rule, is referred to the vicinity of the absçess, but not invariably so. Macewen's symptom (as described in Chapter XXVI.) may be of service, but should be utilized with great caution. The temperature is often lowered in abscess of the brain, and is accompanied by slow pulse. (Macewen.)

Optic neuritis, so characteristic of tumor, also occurs in abscess of the brain. It is not as constant a symptom in the latter as in the former condition; according to Okada's statistics (quoted by Oppenheim) optic neuritis occurred in two-thirds of the cases. The neuritis is in nowise to be distinguished from the neuritis associated with ordinary tumors within the cranial cavity. The other symptoms will vary very much according to the location of the abscess. Paralysis, if present, is apt to be hemiplegic. If associated with unilateral convulsions it points to the motor cortex as the site of the abscess. Sensation is, as a rule, not involved,

and the various cranial nerves are not affected unless the abscess is at or near the base. The fifth and sixth nerves may be pressed upon by a cerebellar abscess, causing pain in the face and paralysis of the muscles of mastication and of the rectus externus. Cerebral abscess may also be associated with paralysis of the facial nerve, but this is only rarely due to involvement of the nerve as it emerges from the pons, but is more frequently of old standing and due to an early disease of the ear affecting the nerve in its course through the Fallopian canal. Paraphasia, amnesic aphasia, alexia, agraphia, accompany abscess involving parts connected with the function of speech. In several patients whom I had the privilege of seeing with Gruening, sensory aphasia was the first signal symptom of an abscess in the temporo-sphenoidal lobe. Hemianopsia may be present (case of Knapp). Stupor, delirium, and coma mark the terminal stages of abscess. In cerebellar abscess the characteristic signs are: Pain over the occiput and the region of the neck; rigidity of the neck; vertigo, inco-ordination and typical cerebellar ataxia; vertigo and vomiting when the position of the head is changed (Bruns); bulbar symptoms.

Differential Diagnosis.—From the account of the symptoms accompanying abscess of the brain it is evident that it is difficult, sometimes impossible, to differentiate between a pus cavity in the brain and other conditions. Among these are solid tumor of the brain, meningitis, and meningo-encephalitis. The diagnosis of solid tumor can more readily be made if the usual causes for abscess, such as ear disease and external injury, are wanting, and if the symptoms have developed in a subacute fashion, which is rather characteristic of tumor, while in abscess there is either a prolonged latent period or all the symptoms develop in a very short period of time. Suspicion of abscess is strengthened above all things by the presence of marked rigors and rapidly changing temperatures. A slight change of temperature may occur in the case of tumors, but it is not as variable and does not show the extremes so common in abscess. During the past years I have observed several cases of abscess in which there was not the slightest rise of temperature for several weeks preceding death.

The differential diagnosis between tumor and abscess is one of practical importance. If the new-growth is near the surface, and if the symptoms are of such a kind as to lead one to infer a tumor of considerable extent, the surgeon might well hesitate to operate if he thought the new-growth a solid one. If there is a probability of an abscess the size of the abscess need not constitute a contraindication to operation, and this differentiation between tumor and abscess is still more important if the growth is supposed to be in the interior of the brain. If this should be a solid tumor a surgical operation would do no good, but if it is an abscess it could well be drained through the hemispheres, and the greatest danger of all, rupture into the ventricles, might be averted.

The differentiation between meningitis and abscess is not an easy one, particularly in cases of ear disease and external injuries. Meningitis is also apt to be associated with, or to precede, the formation of abscess, and during this period the question whether the suppurative, and hence the operative, stage has been reached is not always easy to decide. Moreover, in the final stages of abscess, particularly if the abscess has ruptured, a general purulent meningitis is the invariable result. At that stage a differential diagnosis can easily be made with a great degree of certainty, but if made has little practical value. In the earlier stages of cerebral disease after otitis or external injuries it is safe to believe that the condition is still one of meningitis, without formation of abscess, if the symptoms are those of a general cerebral disturbance, such as vertigo, headaches, nausea, slight stupor, but without rigors or serious rise of temperature. Localizing symptoms may be present in both cases.

The distinction between meningitis and abscess is of much practical importance for another reason. The majority of surgeons would not be willing to operate upon cases of meningitis; they evidently have the general purulent meningitis in mind; with improved methods in surgery, exploratory trephining may be warranted, particularly in connection with ear disease or external injury if there is a suspicion of meningitis alone, provided the latter be limited in extent. Lumbar puncture may give diagnostic aid.

In practice the differential diagnosis most frequently to be established is between abscess and sinus thrombosis after middle-ear disease. The general cerebral symptoms may be similar in both conditions, but the special symptoms of sinus thrombosis (see p. 498) will help to decide the point. In many cases the condition is uncertain before the operation is undertaken. In every doubtful case the surgeon should look first for sinus thrombosis, and then for abscess.

PROGNOSIS.—The prognosis of abscess of the brain is invariably grave. While admitting the possibility of an abcess becoming encapsulated and of remaining latent in the brain for many years, the probability is in every instance that serious mischief will result from it sooner or later. The physician must not trust to good fortune in such diseases, but knowing the serious nature of the trouble he is

bound to present to the patient, or the patient's relatives, the alternatives of surgical interference or death within a limited period of time. The longer the abscess has lasted the more serious it is apt to be, for the danger of rupture becomes greater with the increase in its size.

TREATMENT.—In spite of recent advances in cranial surgery and of the successes that have been recorded in the treatment of abscess of the brain it is best to discuss the prophylactic measures which may possibly prevent the formation of abscess. Knowing that pus is formed most frequently after ear disease and after disease of the bone, the greatest caution should be exercised to prevent any extension of the inflammatory process into the cranial cavity from these original sources of infection. The possibility of cerebral complications in every case of suppurative ear disease should be kept well in mind, and free discharge outward should be secured and maintained. In the case of mastoid and middle-ear disease we are not only bound to secure a free passage outward for the pus, but the discharge should be watched, and as soon as the pus ceases to flow outward while the symptoms point to a constant generation of pus in the bony structures, surgical measures should be resorted to to drain away the pus that is formed. While pus is discharging freely from the internal or the middle ear, if symptoms arise pointing to meningeal or cerebral involvement, such as increasing headaches, vertigo, vomiting, and fever, further surgical aid is needed.

An attempt may be made when the first meningeal symptoms arise to treat these in the ordinary way by the application of cold, by blistering, by counter-irritation, and the application of leeches and the like, but very little time should be wasted with such measures, and if the symptoms do not yield within twenty-four or forty-eight hours the surgeon must afford the needed relief. In the case of abscess due to ear disease, until recently little success has attended the efforts of the surgeon to locate and drain the abscess; but it bids fair to become the most satisfactory and legitimate cause for operation upon the brain. This opinion (of the first edition) has received fullest indorsement by the experience of recent years.

The temporal convolutions can be exposed by trephining at a point which is reached by drawing a line one and a quarter inch back from the external auditory meatus, and drawing another, at right angles from this one, the same distance upward. The terminating point of this line may be made the centre of the trephine opening, and will be sufficiently accurate to enable the surgeon, after removing a considerable portion of the bone, to expose the region of the abscess. As the abscess is not always superficial, punctures should be made with a fine hypodermic needle in various directions in order to reach cavities that are situated below the gray matter. By observing these rules one can hardly fail to find the abscess. In one case of the author's, the surgeon had to make four punctures into the substance of the brain before pus was withdrawn. Abscesses formed in connection with disease of other parts of the cranium can generally be found if one is guided by the external signs; thus, in a case which was operated on for me nearly ten years ago, there was caries in the middle of the parietal bone. The surgeon was directed to open the skull at this point, and, if no abscess presented, to insert the needle in various directions. When the dura was exposed nothing abnormal was noticed, but the first thrust of the needle secured a large quantity of pus; the brain tissue was then incised and the entire abscess cavity opened, but the patient died a few days later, and on the post-mortem table other abscesses were found in the opposite hemisphere of the brain as well as at the base. If *multiple* abscesses can be diagnosticated, operative interference is useless. In cases of skull injury the trephine opening should always be made at the seat of the external injury, and enlarged from this point to meet

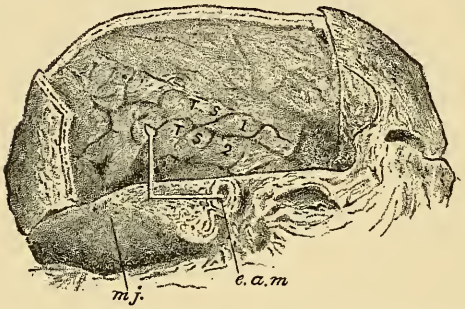


FIG. 123.—Dissections showing the Guide Adopted by Barker in Successful Trephining for Abscess from Ear Disease. (After Gowers.)

the exigencies of a given case. If the pus cavity be due to disease of the nose, a large opening in the frontal bone should be made, from which the surgeon will then attempt to locate the exact seat of the abscess, and, if possible, drain it according to the usual method.

Abscesses in the cerebellum are now within reach of the surgeon's knife. Koerner has collected 55 cases of cerebellar abscess, with a favorable result in 52.8 per cent. of the cases. It is well, however, to remember that the majority of unsuccessful cases are not recorded.

Successful treatment of brain abscess has been reported by Stimson, Bergmann, Schede, Paget, Park, Koerte, Jansen, Knapp, Gruening, Starr, Saenger and a host of others. The case of Knapp is of special interest, as it occurred in a child nine years of age. The chief symptoms were chronic otorrhœa (after scarlet fever), optic neuritis, homonymous hemianopsia, and Macewen's symptom (percussion note stronger on left side); there was no sensitiveness on percussion of the skull. At the operation a pus cavity was found, and complete recovery ensued in the course of several weeks, after minor mishaps.

There is no reason why children who present indubitable signs of brain abscess should not be operated upon according to the same principles which hold good in adults.* It is probable that with greater experience in these cases the operation need not last several hours, as in some of those hitherto reported, and hernia cerebri can surely be avoided.

THROMBOSIS OF THE INTRA-CRANIAL SINUSES.

The blood circulates in the venous sinuses within the skull under special disadvantages. The walls of the sinuses are rigid, the blood cannot be emptied during inspiration, and many of the cerebral veins join the longitudinal sinus at an obtuse or right angle, so that their blood is poured into the superior sinus in a direction opposite to the main current. Under such conditions it is natural that the current should be retarded and coagulation of the blood easily set in.

Two forms of thrombosis occur; the first is the primary or marasmic thrombosis which occurs in children after exhausting diseases (cholera infantum, etc.). This is generally confined to the longitudinal sinus, but may involve the lateral and cavernous sinuses. In such cases the clots are dense, resistant, organized, and do not adhere to the wall of the vein. The throm-

* The readiness of the aural surgeons to open up the mastoid will do much to diminish the frequency of brain abscess.

bosis may be limited to the cerebral veins, thus giving rise to limited or cortical symptoms. Gowers, it will be remembered, has attached special significance to this condition in the causation of infantile cerebral palsies.

The second form is the secondary or infective thrombosis, which generally affects the lateral, the cavernous, or the transverse sinuses. It is secondary to some infective process in the neighboring tissues or at a distance. Middle-ear disease is the most frequent cause, but it may also be due to some traumatism of the skull (infected wounds), erysipelas of the head and face, to purulent disease of the eyes and of the nose. In one of the author's cases caries of the jaw was the starting-point of sinus thrombosis. In other

FIG. 124.—Girl, Aged Twenty. Exophthalmus, with internal strabismus; œdema of the right eyelids, side of the nose, the brow, and the face. Later in the disease the left side was also affected. Thrombosis of cavernous sinus. (Macewen)

cases the infectious material is carried along the veins and through neighboring tissues.

The inflammation may extend directly to the walls of the sinus and thus cause clotting of the blood within, or the clot may form within a vein and extend from there into the sinus. The superior petrosal and the lateral sinuses receive their blood from the middle ear, hence the frequency of thrombosis of these sinuses in middle-ear disease. Thrombosis of the sinus resulting from actual compression is very rare indeed and of little significance, as the symptoms would be obscured by those of the primary affection.

SYMPTOMS.—The symptoms of sinus thrombosis are often complicated by those of the primary disease. We must distinguish between general and special symptoms. Among the former manifestations the most important are intense headaches, somnolence increasing to stupor and coma, convulsions, slight rigidity of the neck, optic neuritis, rigors, accelerated or diminished

pulse-rate, thready pulse, and fluctuating temperatures; but most of these symptoms would suggest meningitis or encephalitis quite as readily, if the conditions favoring sinus thrombosis were not known to be present; moreover, actual meningitis may be caused by thrombosis of the sinus, particularly if such thrombosis is of an infective nature.

The *special* symptoms refer particularly to disturbances in the venous circulation and to areas of tenderness. Thrombosis of the superior longitudinal sinus is characterized by œdema of the scalp and distention of the veins in the parietal, frontal, and occipital regions. In infants the anterior fontanelle becomes prominent, and epistaxis is frequent; squint is a rare occurrence. Cavernous sinus thrombosis is rare (the author has seen two cases within a single year); the thrombosis is generally due to extension from other sinuses, from the petrosal or lateral sinus, or it may result from disease of the ophthalmic veins (Knapp) and abscess in the orbital cavity. In more than half of the cases the thrombosis spreads to the cavernous sinus of the opposite side. The special symptoms are exophthalmus, œdema of the eyelids and root of the nose due to venous obstruction; also ptosis, paralysis of rectus externus, and supraorbital pain, all due to compression of various nerves to the orbit running close to the sinus. (See Fig. 124a.) "Choked disk" from obstruction of the retinal and ophthalmic vessels is present in some cases of cavernous sinus thrombosis. Defect of vision and amaurosis may be superadded. Some of these cases live five or nine months, but all end fatally. The author had a case under observation in which the condition has remained practically unchanged for nearly two months.

Thromboses of the petrosal and lateral sinuses are the most frequent, and the most important; unfortunately they present very few special symptoms. In lateral sinus thrombosis there may be distention of veins and œdema over the mastoid process, and tenderness on percussion in this region, but the symptoms differ very little from those due to ear disease and localized meningitis, and all three conditions may be present. Inflammation of the veins leading from the lateral sinus occurs in cases of infective thrombosis. When these veins are inflamed pain may be elicited along the internal jugular vein, and over the upper third of the posterior cervical triangle. A difference in the sensitiveness of the two sides will be strong corroborative evidence of suspected thrombosis. Cervical abscess and enlargement of cervical glands may result from infective external thrombosis of the lateral sinus.

The prognosis of sinus thrombosis is always grave. A fatal issue is to be expected, although death may be delayed for a period of several months. The only hope lies in successful surgical interference and in early preventive measures. A few recoveries are on record. The lateral and transverse sinuses have been opened successfully and the clots removed.* Ballance, Sulzer, Schwartze, Parkin, Horsley, Macewen, Bacon, Pritchard, and Cheadle have reported successful cases. Parkin's and Cheadle's patients were respectively nine and thirteen years of age. Henschen has collected 145 cases of

* The details of the operation should be studied in Macewen's treatise, page 307 et seq.

operation for sinus phlebitis following otitis; of these, 85 were cured. Tying the jugular vein before removing the clot seems to exert a favorable influence upon the percentage of cures. Koerner states that there was recovery in seventy-five per cent. of the cases if the jugular vein was tied, and only fifty per cent. recovered if the vein was not tied. Jansen does not favor this procedure in all cases.

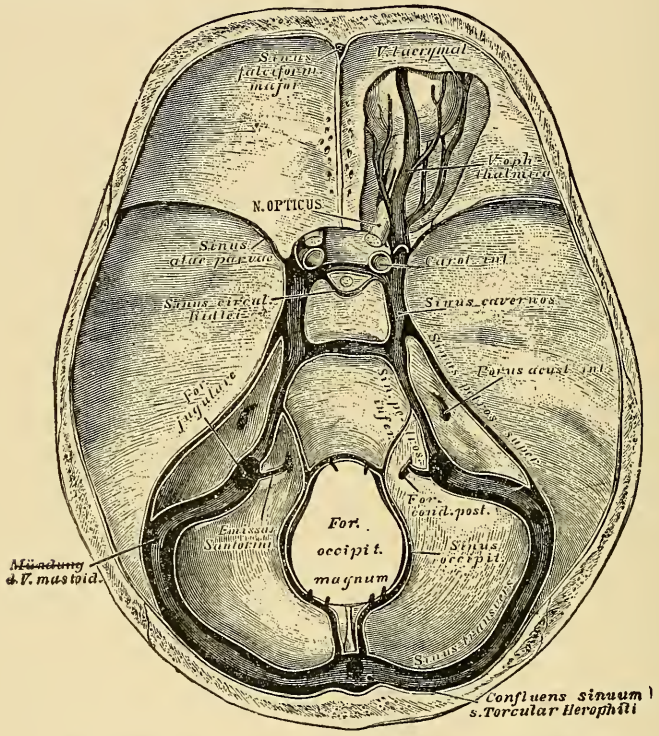


FIG. 124a.—The Veins of the Dura Mater. (Heitzman.)

The extensive experience of aurists and a far larger collection of cases have helped to decide many mooted points, and the successful operations recorded encourage one to hope that sinus thrombosis (excepting possibly as part of a general pyæmia) will become an eminently curable disease. Exploratory operations in doubtful cases are not only justifiable but imperatively demanded.

CHAPTER XXVIII.

DISEASES AND CONDITIONS DUE TO DEFECTIVE DEVELOPMENT OF THE BRAIN.

LARGE DEFECTS.

JUST as there are abnormalities of the spinal cord, due to defective closure of the vertebral arches, so there are deformities of the brain and its coverings due to defective closure of the skull. The various forms of protrusion are best illustrated by Figs. 125–127, taken from Holt's Textbook. These de-

FIG. 125.—Meningocele.

FIG. 126.—Encephalocele. (After Holt.)

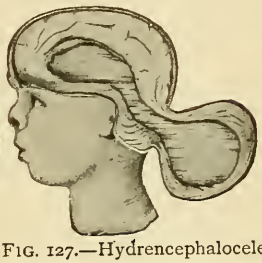


FIG. 127.—Hydrencephalocele.

formities are rare and of little interest, although Treves and others have attempted to remedy these conditions by surgical means. Most of the children thus afflicted die within a few weeks.

In meningocele, the membranes only are protruded; in encephalocele some brain substance is protruded; in hydrencephalocele the protruding brain substance contains a cavity which is in direct communication with the distended lateral ventricle. These tumors, varying much in size, may be occipital or frontal; they may be covered by the scalp, or only by granulation tissue; other deformities are often associated with them.

Cyclops or *monopus*, as the name tells, indicates a condition in which but one orbit exists in the middle of the face, and about in the position generally occupied by the root of the nose. The size of this one orbital cavity varies according to the development of the eye. In some cases the eye is not present, and the orbital cavity is correspondingly small. The cavity attains its largest size if the two eyes are fused into one; and in accordance with the single or double development of the eye, one or two optic nerves may be present. If no bulb has been developed there is also a corresponding defect of the optic nerve. The eyelids may be single or double, the nose, as a

rule, is undeveloped, and its place indicated by something which simulates a short snout. The entire features are distorted, the mouth may be wanting, and the ears come closely together.

The only reason that we have to consider this condition at all is that it is due to a defective development of the anterior cerebral vesicle; the ganglia, too, are more or less deficient; the corpus callosum may be wanting altogether. The parts of the brain coming from the middle and hind brain vesicle are, as a rule, better developed than the hemispheres are.

Fortunately these cases are not viable; they have therefore no clinical value whatever, and may well be relegated to the authors specially interested in monstrosities.

The next most frequent and most complete anomaly of brain structure is the condition known as anencephalus or hemicephalus. The difference existing between the two conditions consists, as the names indicate, in the complete or partial absence of brain development. Steffen assumes, without decided evidence, that these departures from normal development are due to an inflammation of the meninges occurring during the fœtal period, in consequence of which a large amount of cerebro-spinal fluid collects in the ventricles, in the meshes of the pia, and in the subdural spaces; and that in consequence of this increased pressure upon the developing parts the normal growth of the brain is interfered with. There is no reason to dispute this view until some better opinion is offered. The only suggestion to be made is that the inflammation may have been preceded by disturbances of circulation, and if such disturbance of circulation is the important factor, we shall have the same cause operative in these cases of larger defects as in those smaller defects called porencephaly, which we shall study hereafter. In cases of anencephaly or hemicephaly a very considerable portion of the skull may be absent. There may be rudiments of the parietal, of the temporal, or of the occipital bones. In some cases a portion of the occipital bone is left, and this, together with the petrous portion of the temporal bone, provides a small groove in which some cerebellar rudiments may be lodged. The frontal bone is, as a rule, undeveloped. If present, it is very low, the eyes therefore protrude, the face points upward, and the jaws push forward. Instead of the roof of the skull, a thickened membrane may be all that is present. In other cases the bony defect may be covered by a tolerably normal scalp, which may be fused with the dura mater.

If the skull is opened every conceivable defect may be found. In the majority of cases after removal of the bone, the thickened membranes present themselves, and if these are punctured a very large amount of fluid flows out, the membranes collapse, and further examination of the specimen may show nothing excepting the basilar structures, no trace of the hemispheres or any part of the frontal brain being present.

PORENCEPHALY.

The condition of porencephaly is much more interesting from every point of view than the larger brain defects previously mentioned. The term por-

encephaly has been much abused. It has been made to embrace an entire absence of brain structure, or very minute defects in the brain, secondary to vascular lesions. It would be best to use the term in the way it was first used by Heschl, who invented it, and by Kundrat, who wrote the first extensive monograph on the subject. As the name indicates, it is practically a hole in the brain, not an entire absence of brain structure. For that reason there should be some evidence of development of that part of the brain in which the hole lies. This hole, or porus, may be the result of defective development, or it may be acquired in the latest period of fœtal life, or even in the first period of life after birth. It would be well to distinguish the truly congenital cases from the acquired cases, with the additional qualification that even the truly congenital cases may be acquired in the sense of being the result of some morbid process during the fœtal period.

CONGENITAL PORENCEPHALY.—The defect may be in the anterior, in the middle, the posterior, or, in fact, in any part of the brain, but it is found much more frequently in the anterior and in the middle portion. Congenital porencephaly may be single or double.

An extreme condition of porencephaly will be best illustrated by Figs. 128 and 129, taken from Schultze's monograph, which he entitles "A Contribution to the Doctrine of Congenital Cerebral Defects." I select this case because it represents, as it seems to me, an extreme condition of porencephaly, and is only one stage short of a defective development involving a very large portion of the entire brain. By reference to the figures it will be seen that the posterior half of the brain is tolerably well developed, but that the anterior half is practically nothing but a sac enclosed by a thick membrane. The defect included not only the two frontal and a considerable portion of both parietal lobes, but the greater part also of both temporal lobes. The posterior half of the brain, the cerebellum, and all the ganglia proved normal on microscopical examination. Schultze describes this brain very thoroughly, and it is of especial interest inasmuch as its bearer lived to the age of five years. As far as any history may be depended upon, it was simply this, that the child appeared normal at birth, and was supposed to have moved its hands; but this statement Schultze questions, and states that later on both arms were in a condition of flexion and adduction. The legs were flexed, and were in a state of spastic contracture and adduction. The child never had any convulsions, but was able to scream and to suckle. The father of the child noticed that there was no sign of intelligence, that the child would not take hold of anything, that it did not hear, and that it lay constantly with its eyes rolling about, and with its head retracted. The condition was, therefore, one of complete idiocy, with paralysis and contractures of both upper and lower extremities. The child grew weaker and weaker, and finally died of a

bronchitis. The parents of this child had six others, all of them healthy. The mother of the child was addicted to drinking.

Schultze disputes the correctness of Kundrat's views, that porencephaly is the result of anæmic necrosis. This author insisted on the marked distinction between the congenital and acquired cases of porencephaly. If the porencephaly is of intra-uterine origin, the convolutions are so arranged as to radiate toward the margin, or toward the middle of the defect. If the defect is an irregular one, and the convolutions are not arrayed in any definite fashion the porencephaly is acquired after birth. In

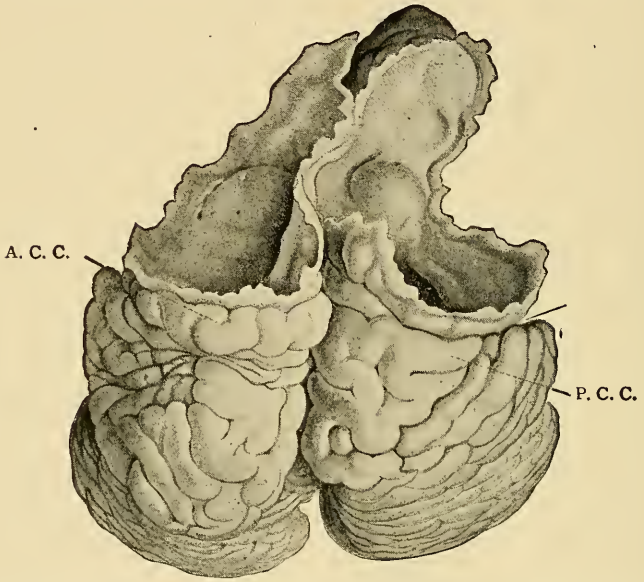


FIG. 128.—Brain with Large Anterior Defect. A. C. C., anterior central convolutions; P. C. C., posterior central convolutions. (Schultze.)

this question, as in so many others, there is some truth in views that appear to be distinctly divergent. I have no doubt that some cases of porencephaly are due to vascular lesions, and others to inflammatory processes which may possibly start from the meninges. The symmetrical defect in Schultze's case seems to me more likely to be due to an inflammatory meningeal process without exudation, at a relatively early stage of fœtal development, than to a destruction secondary to vascular disease. In Schultze's case, moreover, the vascular apparatus was found to be entirely normal. The question arises, What the causes of an intra-uterine inflammatory process could possibly be? If the mother had passed through any pyæmic condition, or had had some form of purulent disease during pregnancy the cause of such inflammatory meningitis or encephalitis would not be far to seek; but in Schultze's, as in

other cases, no such cause could be made out, and it will not do to speculate upon improbabilities. The readiest explanation is to assume, as many of the older French writers assumed with regard to various brain defects, that traumatism plays a very important *rôle*, and that traumatic encephalitis during the intra-uterine period may be the actual cause of some of these porencephalic conditions. Schultze mentions that spasmodic contractions of the uterus without any extraneous disturbance may be a possible cause of such traumatic changes in the fœtal brain. In his own case Schultze is inclined to consider the possibility of a degeneration or obliteration of some of the

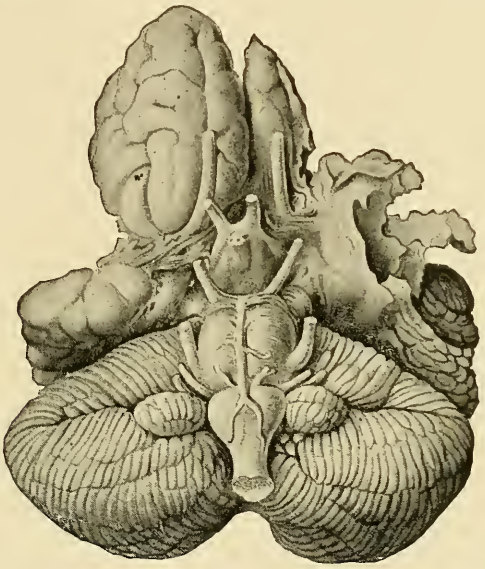


FIG. 129.—Same Brain as in Preceding Figure, Viewed from the Base. (Schultze.)

blood-vessels, or even of an encephalitis, as a direct result of alcoholic tendencies of the mother. In the most recently reported cases of porencephaly of the parietal region there seems to be atrophy of all motor fibres from the cortex to the medulla oblongata. (In eleven of fourteen cases.—Anton.)

ACQUIRED PORENCEPHALY is a condition due to early meningeal hemorrhage. This in turn is the result of compression of the skull during labor. The symptoms associated with these brain conditions will be found described under the heading of Cerebral Birth Palsies. The meningeal hemorrhage is in many cases most severe under the parietal eminences, and over the motor areas. For this reason double spastic cerebral palsy is a natural result of a defect which develops in the region occupied by the motor centres. This region is also supplied by the middle cerebral artery, which is most liable to all vascular disease. The porencephalic defect due to men-

ingeal hemorrhage may be present in both hemispheres, or it may exist in only one-half of the brain. The meningeal hemorrhage, if sufficiently severe, leads to a compression of the cortical substance; atrophy of the cortex follows, upon this the substance shrinks considerably, and the cerebro-spinal fluid is increased either as a result of inflammatory action, or of an effort at compensation. The pia and the cortex are apt to become agglutinated, and thus local cystic conditions with extreme loss of substance may readily occur, the cyst occupying the place which should be filled by normal brain substance.

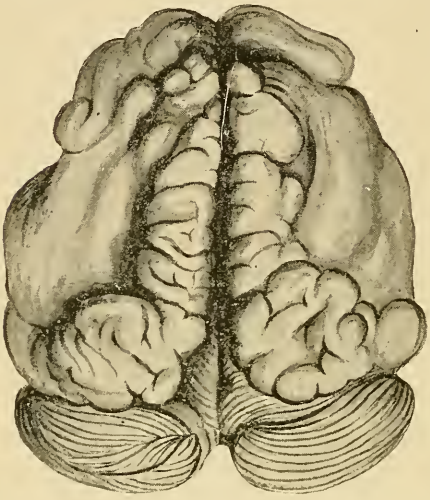


FIG. 130.—Large Double Porencephalic Defect. Child lived to age of twenty months. (From a brain kindly furnished by Dr. Peterson.)

Through the kindness of Dr. Peterson the author is enabled to insert an illustration (Fig. 130) of the brain of a child that died at the age of twenty months. The child had been subject to convulsions and had presented complete spastic paralysis of all four extremities; the head measurements were extremely small (circ. 32.5 ctm.). Over each hemisphere a large portion of the convolutions was wanting. The vacuum was filled partly by subdural fluid, and partly by the bulging of each ventricle, for there was also internal hydrocephalus. Peterson thinks the condition due to a double meningeal hemorrhage, but the defect corresponds so nearly to the distribution of the middle cerebral arteries that obliteration of these blood-vessels early in fœtal life may have been the prime cause.

MICROCEPHALUS.

In addition to the anomalies of brain structure in which one or more parts may be defective, we have to consider that condition in which the brain as a whole shows a defective development, failing to attain to the normal size. There is, as a rule, a correspondingly small development of the skull, and to this small head the term "microcephalus" is given. While a microcephalus is bound to harbor an abnormally small brain, we must not forget that a skull of

normal size may also contain a small brain, the cranial cavity under such conditions being filled up by an excess of fluid. Microcephalus is evidently the result of a number of different processes. It has been a habit to explain it altogether on Virchow's theory, that the smallness of the skull was due to a premature ossification of the cranial sutures, which interfered with the proper expansion of the skull, and that in consequence of this stunted growth of the skull the growth of the brain was arrested. This explanation will hold good for a considerable number of cases, but I am firmly convinced not for all. We must allow also for those cases of microcephalus due to defective cerebral development during the intra-uterine or during the early extra-uterine period. For reasons which we cannot always fathom, the brain does not grow and the skull remains small. The children are small-skulled and small-brained. In another series of cases the microcephalus is, I am convinced, the result of early vascular lesions. I have in mind several cases of children who were born normally, whose cerebral capacity was on a par with that of other children up to the age of six, seven, or nine months. Then convulsions set in, probably resulting in extensive meningeal hemorrhage, or, let us say, in an extensive pachymeningitis hemorrhagica. In consequence of this the cortex was compressed; it became atrophic and sclerotic and, remaining small, the skull did not expand in normal fashion. Such children become microcephalic after the first year of life, or later.

This leads us to a question which has been much discussed and unsatisfactorily answered, whether the growth of the skull depends upon the growth of the brain or *vice versa* (Meynert, Virchow, Lucæ). The two hold a very close relation to one another. If premature stenosis sets in it is unquestionably sufficient to account for a small brain; but, on the other hand, a growing brain evidently helps to expand a normal skull. The fact that defective brains are often covered by normal skulls if the defect has been compensated by a local accumulation of fluid, would seem to prove that as long as the same mechanical conditions are present the growth of the skull remains tolerably normal (Gudden).

In order that we may be able to determine when a skull may be termed microcephalic, we must refer to the meas-

urements of normal heads at certain ages (see Introductory Chapter). If a head which is supposed to be microcephalic fall far short of such average measurements the diagnosis of microcephalus may be safely made; but the head may be developed in proportion to the rest of the body; a small measurement in a child need not necessarily imply a condition of microcephalus, unless the measurement of the skull is below, while other measurements of the body are fully up to, the average in other children.

The general measurements of the skull are not always sufficient to prove whether the brain is small or not. Each part of the skull should be inspected *per se*. Thus I have seen extremely small and receding foreheads with unusually large occiputs. This would tend to give a measurement of a general circumference quite up to the standard, yet on closer inspection there would be no doubt that the frontal portions of the skull and of the brain were poorly developed, whereas the occipital portions were either normally developed or caused to bulge by the accumulation of hydrocephalic fluid within the ventricles.

In several other heads which the author has examined this defect in the anterior portion was extremely marked, and as the frontal portion of the brain has least to do with motor and sensory functions, and more with the general intellectual development of the child, we can see the important bearing such facts have upon the prognosis of a child's future mental development. A frontal microcephalus may be present even though the cranial measurements be up to the average.

The symptoms due to microcephalus are described under the heading of idiocy, and in the chapter on the Cerebral Palsies of Children.* In this connection we may refer, however, to efforts made within recent years to correct these cranial deformities on the supposition that if the size of the cavity could be enlarged the brain would have a more favorable opportunity for further growth. Such reasoning could apply merely to those skulls in which the chief trouble lies in the premature synostosis of the sutures. If there is an inherent defect in the development of the brain, or if the brain growth has been checked as the result of acquired disease, such treatment is not applicable. The great difficulty, however, is, that it is not always easy to deter-

* Of course microcephalic heads are at times found in persons of tolerable cerebral development.

mine to which one of these causes the defective cerebral development is due. Lannelongue suggested many years ago that in the case of microcephalic skulls a longitudinal opening be made in one or both halves of the skull, thus practically increasing the size of the intra-cranial cavity, or at least diminishing the pressure within this cavity. My experience has not been at all favorable to Lannelongue's doctrine, and I am willing to say that there is not the least reason to advise or encourage the operation, at least according to the method proposed by Lannelongue. This French surgeon operated upon a number of skulls, and claimed that after operation the children showed some signs of increased intelligence and were greatly benefited by the operation. Other surgeons have had but few favorable results to show, and of the cases operated upon there were not many in which a small brain was due to smallness of skull. I have had three children operated upon, and on one patient two operations were performed. Two patients with microcephalic skulls due to premature ossification of the sutures, died very shortly after the operation. The third case was a child, two years of age, whose skull had evidently undergone a premature synostosis. It survived the operation and showed a little improvement, inasmuch as it began to play with other children in the hospital ward, but during the four months after operation did not learn either to talk, stand, or to walk. The improvement was, in fact, very slight indeed. Wishing to give the method a further test, the child was operated upon a second time, and an opening similar to the first was made in the opposite half of the skull. The child died from "shock" four hours after the operation was finished.

Figure 131 is inserted to show the condition of the skull* after the second operation. It will be seen at first glance that the original opening that was made became very much smaller after four months; that a tense fibrous tissue was formed over this opening, and that therefore four months after the initial operation the brain was not under conditions much more favorable than those which preceded the

* This skull was demonstrated by Dr. A. Jacobi, at the International Congress, held in Rome, 1894. In the last ten years the author has not advised this operation.

first operation. I would therefore conclude from my own experience * that Lannelongue's operation is not likely permanently to increase the intra-cranial capacity, and furthermore, that operations upon young children are extremely

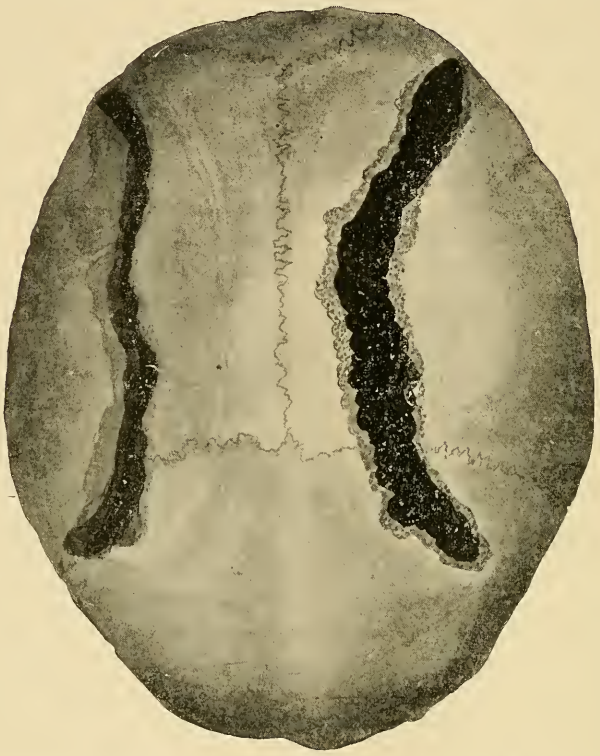


FIG. 131.—Skull of a Child Two Years of Age, showing (on the left of the figure) the Narrowing of the Opening Made According to Lannelongue's Method Four Months Previous to Death. Death ensued four hours after second operation.

dangerous, the children often dying from shock, or from the loss of blood.

If we wish to utilize Lannelongue's idea it will be better to make a circular opening over that part of the brain which from the symptoms in the case would appear to be deficient. In children whose frontal lobes are stunted in growth, circular openings in the frontal or in the parietal bones would give a very large measure of relief, and I am

* Bourneville, Jacobi and almost every one else have come to similar conclusions.

confident that if the operation is entirely aseptic cerebral hernia need not be feared. The bone should not be replaced, but it will be well to provide such children with some form of external protection, or else the danger of direct injury to the brain from falls would be very great indeed. The author wishes once more to emphasize the fact that there are relatively few children in whom a small skull is the primary, or even the most important, cause of arrested development of the brain. The fault lies chiefly with the brain itself and cannot be remedied by operation.

Partial Cerebral Defects.—In speaking of the condition of the brain in microcephalic children I have alluded to the fact that a part of the brain, and not necessarily the entire brain, may be checked in its development. This is particularly marked with regard to the frontal and the occipital lobes. These conditions deserve special attention, moreover, because some of them are attended by symptoms which enable us to diagnosticate during life the exact area of defective development. It is fair to assume that if the frontal lobes are wanting, intelligence will suffer most, whereas the movements of the child will be normal, sensation will be undisturbed, and all the special senses, except possibly the olfactory sense, will be well developed. It is not claimed that intelligence is altogether the property of the frontal lobes, for the various functions that are so essential to normal mental faculties are stored in the most diverse portions of the brain, and a defect in one or more of these sensory divisions of the brain in young children will naturally interfere with normal mental development. If the defect is in the motor areas, the disturbances of motion and of sensation will naturally point out the defective area. While porencephaly is extremely frequent in this region, I have not yet seen a single brain that exhibited normal development of its fissures and convolutions in all but the motor region.

Defective Development of the Occipital Lobe.—Defective development of the occipital lobe, as indicated by an atrophy of the gyri of this lobe, is well illustrated by the case represented in Fig. 132, which has been reported by Peterson. "The patient was a female, twenty-eight years

of age, and was from birth an idiot, with epilepsy and left hemiplegia (slight); she was almost blind, being able only to distinguish light from darkness. Both occipital lobes were greatly atrophied; the loss of tissue was made up for by the widely dilated posterior horns of the ventricles. The cerebellum projected far beyond the posterior lobes;

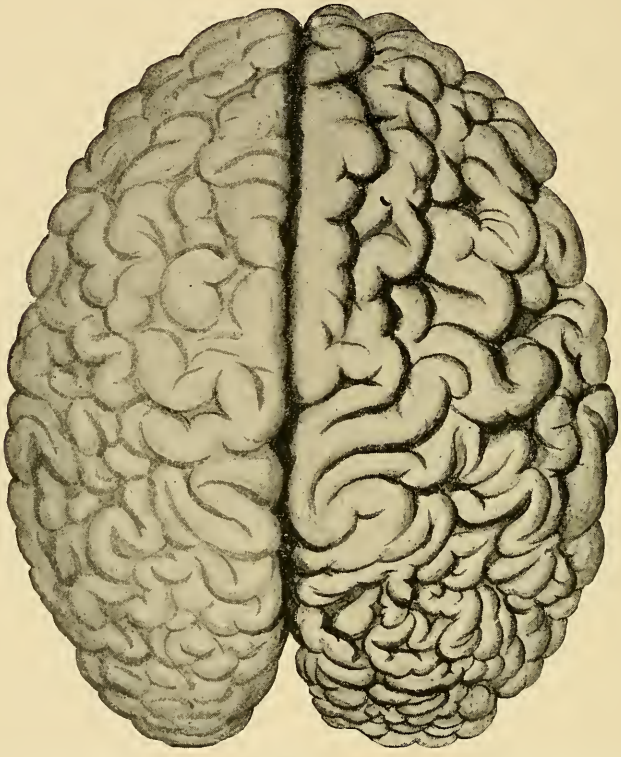


FIG. 132.*—Brain of a Woman Aged Twenty-eight years, Idiot from Birth and almost Blind. Defective development of both occipital lobes, which appear to be mere appendages to the more normal anterior portions. Drawing made from specimen; pia of left half not removed.

the atrophy and sclerosis extended partly into the motor area of the right side."

Defective development of the larger ganglia is not met with in cases in which there are no other large cerebral defects. This statement must be accepted with some reserve,

* The author is indebted to Dr. Peterson for permission to have the specimen copied.

for little attention has been paid to this subject, and of the defective brains that have been examined the records say very little about the ganglionic region.

The author has had occasion to diagnosticate defective development of the cerebellum. The diagnosis was made in a girl, eight years of age, born without difficulty and at full term. The child had never learned to walk or to talk. There was not a sign of spasticity or of actual paralysis. If the girl was placed on her feet she would fall over at once to the right. Equilibrium could not be maintained while the child was in the erect position. Sitting up in bed or on a table she was able to move the various parts of the body. The knee-jerks were absent; hearing and sight normal. There was a condition of complete idiocy. The defect of the cerebellum was evidently part of a general brain defect. The case bore a superficial resemblance to Friedreich's disease, but could be differentiated from it by the early onset and the development of idiocy very soon after birth.

Agenesis Corticalis.—In the conditions previously described gross defects of the entire brain, or of parts of the brain, have been referred to. In the condition of agenesis corticalis the size and weight of the brain may be entirely normal. The changes to be observed are those discovered on microscopical examination. The only outward indication of the imperfect development in the external configuration of the brain is abnormal fissuration. The fissure of Rolando dips straight into the fissure of Sylvius, without leaving a small convolution between the two fissures as in normal brains. (See Fig. 133.) The interparietal fissure extends farther back than normal into the occipital lobe; the island of Reil may be entirely exposed instead of being obscured from view by the margins of the fissure of Sylvius. Such differences as these are characteristic of a lower order of brain development (criminal brains, for instance). On minute examination of such brains that have been hardened, the large nerve-cells in the cortex are seen to be distorted, their contours not as well marked as in the normal brain, the protoplasmic contents are found in various stages of disintegration. The nuclei are either subdivided or destroyed, and the fine nerve-processes, so

beautifully distinct in the normal cells, have disappeared entirely. Sections through the cortical tissue of any part of the hemispheres reveal practically the same condition. The anatomical study of two cases, made some years ago, led to the recognition of a special type of idiocy with blindness and ending in marasmus. (See section on Amaurotic Family Idiocy.) It has been definitely ascertained that the agenesis corticalis is part of a degeneration of the gray matter of the entire central nervous system.

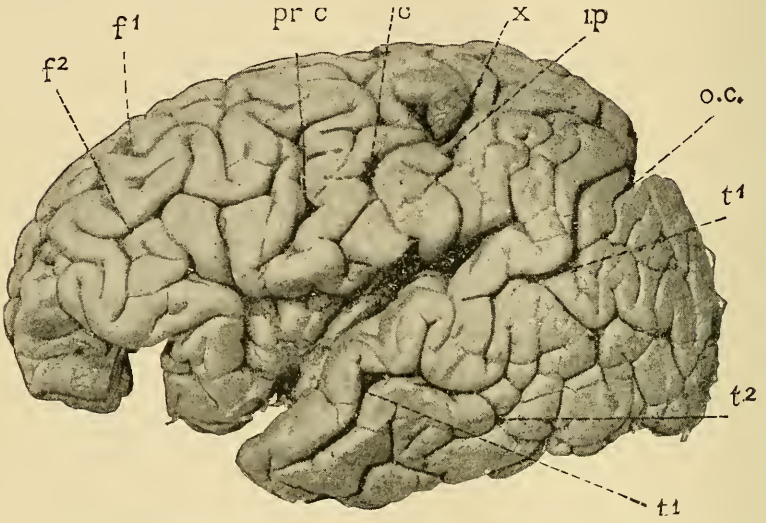


FIG. 133.—Photograph of Brain of Author's First Case of Amaurotic Family Idiocy. The histological condition was described as an "agenesis corticalis." In the above figure, the confluence of fissures and the exposure of the Island of Reil are the signs of a low order of cerebral development. Through the hardening process the conditions have been accentuated a little, but they were present in the fresh specimen. At *x*, and in the frontal lobes, sections had been removed for histological examination. Other letters refer to fissures.

MACROCEPHALUS.

This term may be applied equally well to an increase in the size of the head due to hydrocephalus, as to those rarer conditions in which an increase in the volume of the brain has brought about a corresponding increase in that of the skull. Hypertrophy of the brain is very rare indeed. While some maintain that the increased size of the brain is due to a proliferation of the neuroglia, others believe that it is the result of an increase in all the elementary structures of the brain, but the histological examination has been made in so few cases that the point has not been decided. Steffen

remarks correctly enough that if the increased size were due to a proliferation of the neuroglia, this proliferation would come to a standstill and a partial atrophy and sclerosis would be the natural result, but in those brains which have been examined post mortem true hypertrophy was found and no indications of a sclerotic condition. Rilliet and Barthez, Gerhardt and others, evidently confound cases of interstitial encephalitis with hypertrophy of the brain. Steffen is correct in insisting that the term should be restricted to those in which there is a true hypertrophy of a part of, or of the whole brain. In consequence of the increased growth of the brain the resisting skull causes increased pressure ; as a result of this the blood-vessels are narrowed and the circulation is impaired, whence it follows that the brain is pale and anæmic ; the convolutions are often considerably flattened, and the fissures become more or less indistinct. The ventricles may be greatly compressed, or almost obliterated. By compression from without the cerebro-spinal fluid is driven largely out of the ventricles. As long as the fontanelles remain open the danger from compression is not as great as it is in later years when the entire skull represents an unyielding mass. As long as the fontanelles remain open a true hypertrophy can be differentiated from a chronic hydrocephalus. The hydrocephalus increases more rapidly as a rule than the hypertrophy does. The fontanelle will be more distinctly pulsating in hydrocephalus than in the other condition. The form of the head will vary much in these two processes. In the case of an hypertrophied brain the entire skull will be equally enlarged ; whereas in hydrocephalus the accumulation of fluid in the anterior or the posterior horns of the ventricles tends to a special increase in the frontal and in the occipital portion of the skull. The sutures may be forced apart by the increase in fluid.

The clinical symptoms of this condition are not very different from those found in chronic hydrocephalus ; a considerable mental development, or even normal mental development, is compatible with hypertrophy of the brain tissue, and if the fontanelles are not closed the increase in the size of the brain does not entail a very considerable increase of pressure, but it is false to suppose that children with hypertrophied brains would naturally be brighter than those whose brains are of normal dimensions. The hypertrophy is self-limited by reason of the additional pressure it causes.

If the size of the head increases very much, children are unable to carry the head, feel the need of supporting it, become apathetic, morose, and have a tendency to fall ; if the anæmia increases, convulsions, disturbances of vision, vomiting, and a gradual cessation of all the faculties may precede death, which occurs after an indefinite period of time, unless brought about more rapidly by an intercurrent disease. It will not do to devote too much attention to this condition, which is of extremely little practical value. The diagnosis of hypertrophied brain will be made much more frequently at the post-mortem table than in the wards of a hospital or in private practice.

DEFECTIVE DEVELOPMENT OF THE CRANIAL NERVE NUCLEI should be considered in conjunction with the larger

defects described above. These cases have received but very little attention, yet they are of great interest both from a clinical and from an anatomical stand-point. The nuclei most frequently affected are those connected with the nerves governing the ocular muscles. Cases of congenital ptosis, unilateral or bilateral, are so common that every one can recall persons so affected. Forms of congenital deficiencies in facial innervation come under this heading. Gowers has made a short note of them under the heading of "Infantile Oculo-facial Palsy." Moebius collected over forty cases of this class and reported upon them in the year 1892; while Schapringer gave a most intelligent description of a similar condition which he termed "Congenital Bilateral Pleuroplegia (Paralysis of Lateral Movements) and Facial Palsy." His case, which we shall give somewhat more in detail, shows, however, that his title did not exhaust the clinical symptoms of the case, for there was evidence of involvement of the fifth nerve as well as of the sixth and seventh. I should, therefore, prefer to give to all these diseases the proper designation of "Congenital Nuclear Palsy." That there is in these conditions an actual defect in the development of the cranial nuclei there can be little doubt, and that these defects are associated with other conditions of defective development is proved by the occurrence of these congenital nuclear palsies with imbecility or idiocy. (Oppenheim, Gowers, Heubner, and the author.)

I have seen several interesting cases of congenital nuclear palsy in patients afflicted with scleroderma and myxœdema. We shall be justified, therefore, in relegating to this class of cases all those patients in whom there is a congenital defect of a unilateral or bilateral character implying insufficient innervation of the muscles governed by any of the cranial nerves. The forms that are most frequently observed are congenital ptosis, ophthalmoplegia externa, partial or complete, facial paralysis, and paralysis of the tongue. A close clinical and anatomical relationship would seem to me to exist between these cases of congenital nuclear palsy, and cases of hereditary progressive muscular atrophy, possibly also between these nuclear palsies and those with local muscular defects.

The symptoms of these types will naturally vary very much. I propose giving a short summary of Schapringer's case, which exhibits a more complex association of symptoms than any other with which I have become acquainted. The history in brief is this: A girl, eight years of age (Fig. 134), who was suffering at the time from a slight bronchial trouble, was examined by Dr. Schapringer in 1889. The child was of normal development, and of average intelligence, although she was not sent to school, and had not yet learned to read. In walking she stumbled and fell more frequently than other children of the same age. She states that this is in consequence of a weakness in the legs, and not on account of the visual disturbance. Her face is pale, expressionless, and mask-like. The naso-labial fold is absent on both sides, there is not the slightest indication of folds in the integument of the face either in laughing or crying. Her laughing and crying sounds cannot easily be distinguished. She cannot close her lips. Substitutes linguals for labials, "tata" for "papa," etc. The right angle of the mouth is drawn downward and outward. Whistling, blowing, etc., are impossible. She is not able to wrinkle the forehead either vertically or transversely. When her eyes are open the ordinary amount of eyeball is uncovered. Quite often one lid hangs a little lower than its fellow, but the eyes can be fully and completely opened, proving that both levatores palpebrarum are not involved. Complete closure of the eyes is impossible, although the lids can upon forcible effort be closely approximated to one another. There is a distinct epicanthus, the caruncle and plica semilunaris are only slightly developed on both sides.

FIG. 134.—Girl, Eight Years Old, with Congenital Pleuroplegia. (Schapringer.)

Ordinarily the axes of the eyes are parallel, frequently, however, the right eye is directed a little more upward and slightly outward. She is able to follow an object with both eyes if it be lowered or elevated in the median line. If the object be approached to her face, keeping it in the saggital plane she follows it readily, and she will keep her eyes fixed upon a finger held near the nose for a long period without fatigue. These, however, are all the movements of which the eyeball is capable. The two internal recti contract freely if required in order to bring the eyes into convergence for near objects. If, however, one of these muscles be required to act in unison with the external rectus of the opposite eye for a conjugate lateral vision, there will be found an utter inability to do so. The two external recti muscles are either completely paralyzed or very deficient. The internal recti are not wholly paralyzed. When the child wishes to see an object situated to the side

of the median plane, instead of rotating the eyes she turns her entire head. The fundus appears normal, except that the vessels are a little more tortuous than usual. The functions of the ciliary muscles are normal. There is no strabismus convergens. The size and mobility of the pupils are normal. When the tongue is protruded it appears to the left of the median line; the left half is a trifle smaller than the right. When eating, the child is obliged to use its finger to dislodge food from the cheeks. She is unable to masticate hard substances, such as crusts. She can move the lower jaw laterally toward the right side, but not toward the left; thus indicating a paralysis of the right pterygoid muscle. She has uvula bifida. There is an hypertrophied condition of the canine ligament of the right side, and normal development of the left. In addition to these congenital defects the author mentions a deflection of the distal phalanges of the index-fingers at an angle of about one hundred and fifty degrees in the direction of the middle finger. He interprets this also to be a congenital abnormality. The child has the condition known as "funnel chest."

Reviewing the symptoms, which I have quoted very freely from Dr. Schapringer's paper, it is evident that the congenital defects involve the motor branch of the fifth, the hypoglossal, and the facial nerve, also the tracts which govern the conjugate lateral movements, viz., the third and sixth nerve nuclei, and the nerve-tracts connecting the same. The only criticism to be passed upon this interesting case of Dr. Schapringer's is that while he has invented a good term for the paralysis of the lateral movements, the case presents other symptoms; and that the congenital bilateral pleuroplegia is the most prominent symptom of the condition which we might define in an impartial way as one of bilateral congenital nuclear palsy. Cases of this description are evidently rare, for Schapringer had been able to collect but four others like the one he described.

During the past ten years a number of authors have reported instances of congenital unilateral facial palsy, presumably due to defective development of the nucleus. Schultze described this condition in a girl of five years, Bernhardt in a man of twenty-four years; and Remak has given an account of a young man of eighteen years, who presented a congenital defect of the platysma myoides, associated with slight bilateral ptosis, and a limitation of ocular movements upward. Surgical treatment only is worth considering.

DISEASES OF THE MIND.

CHAPTER XXIX.

INSANITY.

In the preceding chapters we have discussed the organic diseases of the brain, and have touched upon conditions due to arrest of development and to morbid affections of the central nervous system during the period of early growth. Mental disorders may be similarly subdivided. Idiocy represents the psychic derangement due to arrested development; it is so distinct and so large a subject that it will be treated separately. Insanity includes the morbid conditions coming on after a fair degree of mental development has been reached, and constitutes the subject-matter of this chapter.

Insanity in the child resembles closely that in the adult. Such differences as do exist are dependent upon the unusual sensitiveness and irritability of the youthful mind, and upon the paucity of concepts which have been formed prior to the development of the insanity. At a very early day sensory impressions become fixed; hallucinations are possible, therefore, in very young children. But the normal mind does not develop systematized concepts nor the logical faculty during the first few years of life, and prior to this stage of mental development delusions—faulty, unfounded beliefs—cannot exist. The delusions of an insane child will differ from those of the adult in view of the limited ideas it possessed during the period of mental health. Its delusions will be concerned largely with its own individuality and its relation to the family, its teacher, and its God; but the delusions of the adult in regard to his wealth, his position in the religious, social, and political world, are entirely foreign to the child's mind.

The normal child lacks, moreover, the judgment and the power of self-restraint by which the adult is guided and controlled in his actions. Meynert has charmingly depicted the egotism of the child, and the constant struggle of the healthy individual to rid himself of the ideas of self until he

is enabled to sacrifice his individuality and his entire being for the good of the family, the state, and of all mankind. The child for a long time remains upon that lower plane in which passion and the gratification of desires are the mainspring of all action.

The experience of centuries has taught us that by gradual stages, and through the influence of training and education, the selfish views of the child can be transformed into the more generous views of the adult. This process of evolution may be checked by disease; it is only by comparison with the views and attainments of children of the same age that we can establish the standard to which every child should conform.

Insanity in childhood is relatively rare. According to the statistics of Hagen (quoted by Spitzka) only one in 70,684 children annually becomes insane, excluding those born so. Spitzka states that "over four per cent. of 3,244 adult recorded private cases may be justly regarded as having been insane in childhood, while but twelve patients afflicted with infantile forms of insanity" were observed by him during their infancy. From this it is evident that we must not only regard the full-fledged insanities of early life, but also those morbid conditions in early years which foreshadow the outbreak of mental disease in later years.

The psychoses are not developed in very young infants, although Greding reports a child that was a "raving maniac" at birth. The story must be taken with a grain of salt. Rush has noted the occurrence of "mania" at the age of two years, and Sinkler has observed it in children three years old.

After the period of first dentition is past, faulty psychic inheritance may become noticeable; but the age of puberty is the one fraught with greatest danger. The author had under observation various members of a family, all of whom (two boys and three girls) have developed mental disease between the ages of twelve and fifteen years. All classes of the social body seem to be equally prone to mental disease; while idiocy is unquestionably more frequent in the poorer classes than among the well-to-do.

Etiology.—Heredity is the potent factor in the causation of juvenile and adult insanity. There is always the danger of over-estimating the importance of any one factor, but the daily experience of alienists and neurologists helps to enforce this belief. The importance of heredity is im-

pressed upon us, too, by the different course of acquired insanity in persons with or without an hereditary taint. Psychic disorders constitute, as a rule, the last in a series of degenerative diseases; neurasthenia, chorea, epilepsy in the parent may lead to insanity or idiocy in the child. By way of exception healthy individuals may emanate from families with a distinct neurotic or psychic taint. Mental derangement in children is ordinarily developed, if one or both parents have been insane, if one or both have been afflicted with a severe neurosis, if one or both have been addicted to alcohol, and above all, if the parents are related, and belong to a family in which nervous or mental disease has been of common occurrence. Individuals of different but tainted families will also be liable to generate insane children, or such as show distinct stigmata of degeneration (deformities of the skull, of the ears, gothic palate, stunted growth, abnormal sexual appetite, early masturbation, and the like). Closely allied to the hereditary forms of juvenile insanity are those due to traumatism during birth. Meyer has insisted on this. The author has seen innumerable cases of idiocy due to this cause, but not a single instance of any other form of mental derangement.

Arrested physical growth is often associated with mental impairment due to hereditary causes. A girl, aged twenty, whom I have seen in repeated maniacal attacks, is stunted in growth, being only as tall as a child of twelve, without any sign of mammary development or of menstruation. The girl stutters a little, but is fairly bright in the interval between the maniacal attacks.

Among the causes of insanity acquired in early childhood, the chief one is beyond doubt traumatism to the skull. Emminghaus has found this factor in fourteen of one hundred and three cases, not including those in which the injury was followed by epilepsy and mental derangement. Loss of memory and complete dementia, or maniacal excitement, are the forms of insanity commonly developed from this cause. The occurrence of idiocy after relatively slight injuries need not be insisted on in this connection. Meningitis early in life, ear disease, insolation, epilepsy, hysteria, chorea, severe emotional excitement, grief, shame, disease of

the heart, intestinal parasites, acute infectious diseases are the conditions which predispose to the development of insanity. To these might be added excessive work at school, maltreatment at home, starvation, and cruel punishments.

Masturbation is another and a potent cause. The importance of this factor has been underrated by some; it can scarcely be overrated. There is a just difference of opinion, however, as to the influence it exerts in generating mental disease. Many claim that it is often the early symptom of an insane condition. There can be no doubt of this, but it is equally certain that the continuance of the habit brings about a rapid deterioration of the individual's mental state. It is often acquired by imitation, and quite as often it would seem to be the result of natural tendencies. In the former class the practice may occasionally be checked; in the latter, the habit is continued, both in boys and girls, in spite of the most persistent watching. The more stubborn cases are generally those characterized by other distinct signs of degeneration. The evil effects of the vice become most pronounced at the time when the individual is subjected to the first serious strain. Several of such young patients have done well at school, have passed their examinations successfully, but have broken down completely in the first competition with boys of mature and normal intellects.

FORMS OF INSANITY.

A detailed account of all the symptoms of insanity would far exceed the limits of this treatise. By combining a discussion of the symptoms with a description of the types of mental derangement, as they occur in early life, we shall be able to give a sufficient survey of the entire subject. It is best to begin with those symptoms and conditions which are on the border-line between sanity and insanity.

IMPERATIVE CONCEPTS.—The orderly sequence of thought in the child and in the adult may be interrupted by foreign and recurrent concepts, which obtrude themselves upon the individual's consciousness, which annoy him and tyrannize over him, but of which he is not able, at least for a time, to rid himself.

These imperative or insistent concepts are very common in childhood; at times they disappear without leaving any trace of mental defect behind them; in some children they represent the first stages of chronic forms of insanity. Imperative conceptions may lead to imperative actions, to morbid fears, and morbid impulses.

Many normal persons have passed through physiological states akin to these imperative concepts. On going to bed the thought arises suddenly that the light has not been properly turned down, that the gas might be escaping. They assure themselves that all is well, and after a few minutes the same fear or concept comes upon them again. The person so troubled may attempt to allay his fears by methods which ordinarily carry conviction, but in spite of himself the same insistent idea annoys him. Returning to one's door again and again to make sure that it is properly locked; the annoying thought that the letter which has just been mailed was not properly signed or sealed; such are some of the imperative concepts of daily life. The sight of a knife may suggest the harm that can be done with it; a person standing on a precipice or on a high tower has a feeling as though he would be dragged into the depths below. The child, after it has passed the first five or six years of life, may have similar experiences. It is often tortured by these insistent concepts. Some pupils at school have to do everything over and over again; they never finish a sum in arithmetic as quickly as others because the doubt arises whether they have done it properly, and they are impelled to do everything a definite number of times (arithmomania). As a rule, they can assign no reason for this. A little patient would never address any one without repeating the name twice; he was conscious of this peculiarity, was finally induced to repeat the name a second time to himself in such a low tone of voice that no one could hear it, and gradually lost the habit.

Another lad, well known to the author, would do everything three times for fear of having done it twice. He would take three steps at a time in going upstairs; if there were but two he would jump back from the first and then walk up the two, so as to make three in all; before going to bed he touched the floor three times; as soon as he was in bed he would get up again to make sure that he did not touch it twice. These concepts and actions were imperative, but they were developed from a fear that was engendered in him at the time of his father's death that his mother might be the second one to die. This caused him to avoid "two" in every way. After a number of years the imperative concepts were overcome.

Closely allied to these conditions are the fears of contamination (the mysophobia of Hammond; délire du toucher of Legrand du Saulle). I have known several children who would not touch the knob of a door, or take

anything out of other people's hands. One child would not be washed because the water flowed through pipes and the pipes were not clean. The fear of open places (agoraphobia of Westphal) occurs in children, though not nearly so often as in adults. Some years ago the author treated a young girl, twelve years of age, who could not be induced to come to his office because she would have to cross a small hill over a tunnel; she was certain that she could never get across. These fears may increase in such number as to give rise to what Mills calls pantophobia.

Children as well as adults are troubled by insistent questions—generally abstract ones—which interrupt their trains of thought. Why is the sky blue, the water wet, the ball round? Occasionally they soar into the field of religion. Why is God just? and so on.

All these conditions may be of slight significance unless they lead to imperative acts of violence (cutting a child's throat at the sight of a knife, etc.); or unless the effort to suppress these morbid concepts and impulses produces great excitement. A little patient of mine, years ago, was so annoyed that he would every now and then pass into a maniacal state or entertain suicidal ideas. Westphal was of the opinion that these imperative conceptions were never developed into delusions; this is true in general, but they are often present in persons who develop systematized delusions suggested by the contents of some one imperative conception. Imperative concepts and impulses are very apt to occur in children who have passed through some exhausting disease, or some intense emotional excitement. As children grow older they may learn to disregard annoying concepts, and thus rid themselves of them.

Cerebral Neurasthenia. — Nervous exhaustion, or neurasthenia, is, on the whole, so rare in children that we have not devoted any special chapter to the discussion of this disease. The causes which give rise to neurasthenia in the adult are far less potent in childhood. Excitement, over-work, cares in early years, are more apt to produce hysterical and choreic conditions than neurasthenia; occasionally, however, the cerebral type is developed in young persons between the ages of ten and fifteen years or older.

The chief symptoms are, inability to concentrate the attention upon any special work, or excessive restlessness following upon such an effort. A feeling of pressure upon the top of the head, a dragging sensation over the nape of the neck and the upper portion of the spine, general irritability, a morose disposition, sleeplessness and a feeling of fatigue, a slight increase in the reflexes, sensations of heat and cold complete the details of this condition.

Cerebral neurasthenia occurs mainly in the children of hysterical or excessively neurotic parents, and in those who come of healthy parentage but are pushed inordinately in their work at school to satisfy the demands of ambitious parents. It has been the custom to attach the entire blame to present educational methods, but the chief fault lies with the father or mother who cannot recognize the child's inability to cope with other children of the same age. Boys or girls who recognize that they are at a disadvantage in competition with others become discouraged and moody, and often develop a condition of melancholy depression.

Some years since, a young girl, aged thirteen, was brought to me, who had just entered upon the work of a high-school in which an older sister had graduated with high honors. The patient was told by her mother that she would be a discredit to her family unless she did at least as well as the older sister had done. The child made a strenuous effort to come up to these expectations, but the result was evident on examination. The child complained of intense headaches, frontal and vertical, of a feeling as though she were being crushed under a weight. She looked pale and haggard in consequence of the loss of sleep. Her appetite had left her, and for a few days before she was examined was either in a drowsy condition or else would cry and complain of her misfortune. She knew that she could not possibly do satisfactory work at school, and had developed the idea that she was thoroughly unworthy of the care which her parents had bestowed upon her.

The child was taken out of school at once, sent to some relatives in the country, and was told that she would get entirely well provided she would not think of school, or anything connected with school matters, for a period of several months. The parents were also instructed never again to force the child's work, or to foster any inordinate ambition in her. The older sister was a bright, strong, well-developed girl, who could in all probability have stood any amount of work without showing signs of fatigue. The younger child was less well developed physically, and presented the remnants of former internal hydrocephalus (a bulging forehead and very large transverse

occipital diameter). Had the parents been allowed to continue in their foolish course much longer, this child, who presented on the whole none but neurasthenic symptoms, would unquestionably have developed a serious form of psychosis.

The course of cerebral neurasthenia may cover a period of several weeks or months, according to the time at which a radical effort is begun to check the development of the disease; but the affection is an eminently curable one. The condition should be treated very much as is the same disease in the adult. The most important point is to give body and mind complete rest, to take the child from school and to prohibit all mental fatigue either in the way of school or of home studies. If a proper arrangement can be made, it is best to place the child under care of a sensible nurse, and subject it to the rest-cure. A separation from hysterical and irritating relatives is absolutely essential. Careful feeding and the ordinary tonics—above all, iron, arsenic, and small doses of strychnia—will help to bring about a rapid improvement. After four to six weeks the child should be allowed to roam about in the country, and its mind should at least for a year or more be kept entirely free from every sense of duty and obligation.

Hypochondriasis is often associated with neurasthenia, and occasionally it is a primary affection. The fear of insanity is not as pronounced as in adult neurasthenics, but the fear of death is common. The uncomfortable sensations in the head frighten the child, and soon it takes notice of every little symptom to which other children would pay no attention. In such introspection children are encouraged by over-anxious mothers. Like the adult, the young hypochondriac hears his heart palpitate, takes notice of the rumbling of his bowels, watches his skin most carefully, and exaggerates the importance of every pimple that appears upon it. At the age of puberty a boy's attention is rivetted easily upon the sexual organs. He may be frightened by the occurrence of erections or by the difference in the size of the testicles. A bright lad, aged fourteen, whom I treated for a long time, and who came of a highly neurotic family, was at great pains to prove to me that his testicles were "detached" from the rest of his body, be-

cause he could move them about freely. Early masturbation intensifies such fears. Westphal reports the history of a young hypochondriac who complained of abnormal sensations in the head, in the feet, in the abdomen; his tears had been dried up, and all the mucus in the lungs had been expectorated. The boy's hypochondriacal mood was interrupted by a mild attack of maniacal excitement. The prognosis of hypochondriasis in the young is entirely favorable unless it is based upon a marked hereditary taint. The cure of the condition lies in sensible management of the youth's mode of life, in diverting his mind from himself by moderate indulgence in out-door sports, by providing him with the companionship of sober-minded lads of the same age. I have known boys suffering from sexual hypochondriasis to be forced by parents into early intercourse with the opposite sex. The procedure is invariably harmful.

The preceding forms of mental derangement are less serious than those which we must now consider.

Mania.—The term "mania" denotes a form of mental disease in which there is a marked acceleration of all cerebral and physical functions, generally associated with a feeling of well-being. In children this condition is relatively rare; it is often confounded with a temporary active delirium, which comes on after the ordinary febrile diseases of childhood. In true mania restraint is entirely removed; the patient's ability and desire to do anything and everything he chooses knows no bounds. Thoughts and impulses follow rapidly upon one another. Emminghaus has well said that an exalted mood, a rapid succession of incomplete thoughts, unbounded desire to make everything it sees its own, are characteristic of the healthy child. But the normal child, under the restraining influences of education and training, soon learns to curb its desires and to develop an orderly train of thought. The young maniac has apparently cast aside all restraint and gives himself up completely to his passions and his impulses. His incessant activity leads him to tear and destroy everything within his reach; he is cruel to others and does not hesitate to inflict injury upon himself. There is no trace of a sense of decency or propriety, even young children using the foulest language. Immature

thoughts follow rapidly upon one another; everything the maniac hears amuses him; a word suggests another that rhymes with it; his answers are often quick, sometimes bright; but they are flashes merely and do not denote real intelligence.

The excessive restlessness and the rapid flight of ideas are not followed by a feeling of fatigue, which would be natural in a normal person. Loss of appetite and of sleep contribute to the exhaustion which is developed if the condition lasts for a number of weeks or months.

The symptoms of mania come on in an insidious fashion, or may be preceded by a period of depression. At first the greater liveliness and activity of the child are supposed to be a mere exaggeration of its normal state; by degrees the increase of all the symptoms points to a morbid state; after the acme of the disease has been reached the condition remains unchanged for some weeks, and then the excitement gradually diminishes. A return of sleep is, as a rule, the first favorable sign pointing toward recovery. Recurrent mania (several attacks and remissions) has been described. The exact duration of a maniacal attack may vary between five months and a year, but the state of intense excitement is, as a rule, much shorter.

Among the etiological factors the acute febrile conditions and severe strain or emotional excitement are the most potent. The period of beginning menstruation is fraught with the greatest danger. The only cases of true mania which I have seen were in girls at this period, and in a boy, aged fifteen. One of the girls had passed through a series of examinations at school with great credit to herself. On the morning following the last examination she refused to get out of bed, would not take her food, and would not allow herself to be washed. To all questions why she would not do as asked, she mumbled a few inaudible words; soon she became wholly silent, and for a period of four weeks she lay in an absolutely stuporous condition, passing urine and fæces into the bed, and refusing to take nourishment. From this condition she passed quite suddenly into one of most violent mania, in which she tore everything that she could lay hands on, sang and cursed all day long,

spat at every one, tore her own clothes into shreds, and would expose her person before every one. (In health she was a sweet well-behaved child.) The state of maniacal excitement lasted four months, during which time she lost a great deal of flesh. Six months after the first symptoms had appeared, menstruation set in, and from that time onward a rapid and complete recovery ensued. In other cases I have observed a development of mania after the cessation of the menses that had appeared a few times. A close relation between the menstrual flow and these psychic conditions cannot be doubted.

While the prognosis is favorable, careful treatment is necessary in all cases. These children must be guarded by competent nurses; mechanical restraint cannot always be avoided, but it should not be more forcible than necessary. Such patients are generally fit subjects for an asylum, and often do better there than at home, where greater restraint has to be applied. The author has treated such patients, however, in their homes by placing them in large, well-ventilated rooms, away from the rest of the family and under the charge of two or more nurses.

In home and asylum treatment, the hydrobromate of hyoscin (one two-hundredth to one one-hundredth grain three times daily), given by the mouth or hypodermically, is a valuable drug. In addition, sulfonal, trional or veronal, in moderate doses, should be given at night. Bromides are entirely useless, and opiates of little benefit. Prolonged full baths, followed by slightly cooler ablutions of the spine tend to calm the excitement, and sometimes help to induce sleep when all other measures fail.

MELANCHOLIA represents a distinct form of insanity which has been observed in children, particularly between the ages of eight and fifteen years. It is not to be confounded with simple melancholy depression, which may accompany many other morbid mental states. It is as natural for a lunatic who supposes himself to be the victim of persecution, or a target for the raillery of others, to be depressed and melancholy as it would be for a sane person who found that all his efforts to succeed were in vain, and that everything and everybody were against him.

In true melancholia the child is depressed without cause. Experiences which would please others make no impression upon its saddened mind. It does not care to play with other children, whose frolicsome ways are a source of annoyance. Games, books, theatres, have no charm for the melancholy child; it seeks seclusion, sits in a corner by itself all day long; will not speak spontaneously, and, if spoken to, either does not answer at all or replies in monosyllables and with much hesitation. No reason for its moodiness is given; after much questioning it may say that it feels sad, but does not know why. To the questions put to a little patient I received the answer, "I know, I know," accompanied by a nodding of the head. As it was recovering, the child stated that she knew everyone was against her; that her parents did not care for her; that she was to be punished because she had not loved them enough, and because she was so ugly (as a matter of fact she was good-looking). As this same child was taken to the country to hasten convalescence, she said: "I know, I know, you will burn me to ashes."

In contradiction to the acceleration of all mental processes in mania there is a distinct inhibition, a "slowing up" of all mental and physical processes in melancholia. Loss of appetite, restless sleep, slowness of all muscular efforts, obstinate constipation are characteristic features of this disorder. The child may be able to reason a little regarding its unfortunate condition, and self-accusations are abundant. It accuses itself of unkindness toward others, of want of respect and love for its elders; if it has had any religious training it develops the ideas of the "unpardonable sin," and of lack of devotion to God—ideas which play a prominent part in the melancholy of adults. Hallucinations in keeping with the depressed mood, the sight of the devil, of a cruel teacher armed with all sorts of weapons of torture, may increase the depression, or else lead to a condition of excitement or frenzy (melancholia agitata). Unlike the maniac, the melancholy patient directs his frenzied impulses toward his own person. Self-mutilation is common. A young girl, aged thirteen, had to be restrained because she would insist upon tear-

ing out her hair and attempted to cut deep into her skin.

Suicide of children is frequently the result of melancholia. Like the adult, the child seeks to put an end to a life that is so full of trouble and misery. Statistics as to the frequency of suicide in children are rather unsatisfactory; Emminghaus quotes those of Morselli that show the number to be greatest in Denmark and Prussia, and smallest in Belgium and Italy. The author has seen no statistics bearing upon America. In the majority of cases some mental disease is the cause of suicide in the young, and none is more frequent than melancholy. Some suicidal attempts are due to trifling causes—fear of punishment, chagrin over an unmerited rebuke, etc.

Melancholia with stupor (melancholia attonita) is a more extreme form, which is either developed primarily or follows upon a condition of simple melancholy. In this form the child lies in bed absolutely motionless, takes no notice of its surroundings, and cannnot be induced to smile or to say a single word. It reacts but feebly to every form of cutaneous stimulation; it will to a certain degree tolerate pain rather than move. If passive movement is attempted, the limbs will retain the position given them, or the child offers considerable resistance. Urine and fæces are passed into the bed. During this condition of stupor disagreeable hallucinations and terrorizing delusions add to the child's misery.

In the author's experience melancholia is the most frequent form of mental derangement in childhood. Its course varies very much. Some patients get well in a few months, others do not recover for more than a year. In a number of instances a condition of mania follows upon the period of depression. The prognosis is favorable as regards ultimate recovery; it is certain that sixty per cent. of young melancholy subjects get well. As a rule, the prospects of early recovery are better in the agitated form, and less bright in the stuporous form. Melancholy may terminate in dementia, or in death from exhaustion or from suicide.*

The treatment of melancholia is very simple. One or

* The author has not entered upon an enumeration of the morbid anatomy of melancholia or mania, as there is nothing but pure hypothesis to proceed upon. The vascular theories of Meynert have not been corroborated by others.

more competent nurses must be deputed to watch over the child and protect it against all harm. It must be fed carefully with a spoon, and will do best on milk, some cereals, scraped meat, and eggs. The stomach-tube should not be resorted to unless absolutely necessary. Opium in small doses, or hyoscyamia in the agitated forms, are the most useful drugs ; both can be given hypodermically if necessary, but they should be discontinued as soon as practicable. Sulfonal, chloralamid, or trional will help to induce sleep and to reduce mental excitement. As in cases of mania, a warm, full bath is a valuable aid in treatment.

There is no sufficient reason to remove such children from comfortable homes, if isolation at home is possible ; but if the environment of the child is an unfavorable one, the sooner it is taken to an asylum the better it will be.

Periodic and Circular Insanity are very exceptional occurrences in children. The youngest patient of this class the author has seen was a boy, aged eighteen, and all of the patients described by Krafft-Ebing, Jacobi, Kelp, and others have been near or beyond the age of puberty. Periodic insanity consists of successive attacks of mania or of melancholia, followed by a lucid interval of months or years, and then a recurrence of the same conditions. Circular insanity is closely allied to this form. As described by Krafft-Ebing it is distinguished from the ordinary periodic insanity by an alternation of maniacal and melancholy stages followed by a lucid interval, and then a recurrence of derangement in the same or the reverse order as before. The diagnosis of circular insanity can be suspected, but not made, until the patient has passed through several cycles.

The duration of each cycle may vary from several days to several weeks ; but in the author's experience as the disease progresses the lucid periods grow shorter and shorter. The perodic and circular forms of insanity are of the hereditary degenerative type, and the prognosis in them is far less favorable than it would be in ordinary mania or melancholia. During the maniacal or melancholy state the treatment would be similar to that described for each condition. These patients are dangerous to themselves and their surroundings, as they pass quickly from one stage to

another. As soon as the circular and periodic character of the disease is established, it is best to keep such children under the constant observation of an attendant, or else to place them in special institutions.

The author has not departed from the above account as given in the first edition of this book, although he is well aware of the changes which have been brought about in the classification of early mental disorders through the writings of Kraepelin and his followers, who have substituted for periodic and circular insanity the conception of a manic-depressive form of insanity. With the exception of the involution melancholias, Kraepelin would consider all periodic or recurrent forms of mania and melancholia to be merely different manifestations of a single psychosis, viz., manic-depressive insanity. Startling and revolutionary as Kraepelin's doctrines may appear to be, the writer's ample experience would lead him to indorse this view, at least in part, but he must also insist that many a youthful individual may pass through periods of maniacal excitement or of depression without such an individual necessarily being a victim of a circular or manic-depressive psychosis.

DEMENTIA PRÆCOX: Following Kraepelin's suggestions, alienists and neurologists have adopted this unfortunate term, designating thereby an entire series of youthful or adolescent forms of insanity characterized by a distinct tendency to progressive deterioration of mind. French authors, the writer, Dercum, and others have been at some pains to protest against the too liberal use of this term and against the practice of labeling almost every form of youthful insanity as a dementia præcox, for many of these patients never develop a condition of dementia and should be spared the stigma of a name that implies an incurable malady. On the other hand, it is well to admit that mental enfeeblement is a frequent accompaniment of insanity in early life.

Dementia Præcox includes three distinct forms, each one of which may be developed at the age of puberty or even earlier: (1) Hebephrenia; (2) Katatonia; (3) the Paranoid form of Dementia Præcox.

Hebephrenia as described by Kahlbaum and Hecker is characterized by a period of depression, with hallucinations and delusions of a corresponding type; the patients are often conscious of their altered mental state and recognize their inability to vie with others of their age at school or even in sport; they lack the power of concentration and of attention. Their minds are easily diverted from the subject in hand, their memory and judgment become defective, they are indifferent to family and friends. Volubility of speech may for a time conceal a real mental defect. The patients laugh or cry without reason, are apt to repeat high-sounding phrases, their letters are verbose and meaningless, their conduct and manners become affected, silly, theatric, and their moral sense may be more or less impaired. According to Kraepelin's statistics only 8 per cent. of hebephrenic patients have any chances of recovery.

KATATONIA, first described by Kahlbaum, is characterized by states of stupor or excitement associated with conditions of "negativism," of muscular tension, of impulsiveness, with a tendency to automatic and stereotyped ac-

tions, and with automatic repetition of words. All these peculiar mental conditions lead to distinct deterioration within a reasonably short period of time.

The paranoid form of dementia præcox is characterized by the development of an entire series of extravagant, absurd and ever changing delusions, coming on frequently with an initial period of depression and leading within a few years to a condition of undoubted dementia. Its onset early in life and the rapid development of a dementia will help to distinguish it from true paranoia.

If we accept the Kraepelin doctrines in whole or in part, the prognosis of these forms must be given in very guarded fashion. But there seems, to the writer, to be sufficient doubt in many of these cases as to the ultimate outcome to make it incumbent upon the physician to urge that such individuals be separated from the family at once, that they be given for a time, at least, wholesome employment in the country or elsewhere, and that they should not be made asylum inmates until they have proved either that they are incapable of conducting themselves properly when at large, or that their psychic condition is such that they need institution treatment for their own protection and for the improvement of their symptoms.

PARANOIA.*—Primary insanity is a degenerative psychosis of the hereditary order characterized by hallucinations and delusions. The latter are primary symptoms and not secondary to the exalted or depressed mood, as in mania and melancholia. The delusions or "fixed ideas," become systematized, and dominate the mental activity of the individual to such a degree that they become the mainspring of all action. The paranoiac is not amenable to reasoning; and his delusions cannot be dislodged by argument, as happens with the temporary delusions of the sane. He may have one set of delusions or many. The persistence of one or many proves that the entire logical apparatus is out of gear. It is wrong to claim that any person is insane on one point only; he may show his insanity in one direction chiefly, but his mental derangement is as marked as though he had dozens of fixed ideas.

The systematized delusions of paranoia may be divided into two great groups; first, delusions of persecution; and secondly, delusions of grandeur; the latter may be sub-

* The writings of Snell, Westphal, Sander, Krafft-Ebing, and Meynert, have contributed most to an understanding of this subject. Spitzka treats the subject very lucidly under the name "Monomania," preserving an old term, but discarding the doctrine of the "Monomanias" which did so much harm in psychiatry.

divided into the religious, the political, and the erotic type. In this form of insanity, with delusions of persecution, the patient believes himself to be the victim of circumstances. He is made to suffer either for wrongs he has committed or for the envy others feel toward him. He has, as a rule, been morose and exclusive. He feels that he is being observed by others; that every one notices a peculiarity in him; that others can read and control his thoughts; that the newspapers direct their flings at him; when they speak of rascals, of thieves, they mean him. Before long he hears the voices of his enemies who are trying to ferret out his actions; he stops up the key-holes and draws the blinds of his windows. If his neighbors cannot get rid of him as speedily as they wish, they put poison in his food, which he refuses to take. He supposes himself the victim of the police, of socialists, of a religious sect, who will endeavor to influence him by electricity, through the telephone, by hypnotizing him, or by forcing him to inhale all sorts of noxious vapors.

Paranoia with delusions of grandeur includes all those who imagine themselves destined to fulfil some special mission; the political reformers, the Guiteaus, the religious fanatics, the presidents, the emperors and kings, the Goulds and Vanderbilts among the insane, belong to this class chiefly. Guiteau, descended from a father who believed in Mesmerism and in free-love, was particularly fond of reading on religious subjects; he masturbated at an early age and entered the Oneida Community at the age of nineteen; when twenty-four years old he writes to his father saying that he was employed by Jesus Christ & Co. Some paranoiacs exhibit signs of the insane neurosis at a still earlier day.

The children who are exclusive, who never care to play with others, who pray when their comrades frolic about —these are the very ones who develop paranoia or the paranoid form of dementia præcox. Moody, queer, and "cranky," they go along well enough until they have to compete with others in the struggle for existence, or until they are overcome by some severe grief, by strong emotion, by political or religious excitement (the election campaigns, the

revival meetings, and the like); and then delusions, which may have been latent for a long time come to the foreground. The paranoiac comes of neurotic stock, in which insanity, hysteria, epilepsy, and chronic alcoholism have been common occurrences.

Not all children who are morose and exclusive turn into paranoiacs, but it is well to look with suspicion and fear upon youthful prodigies, who discuss philosophy, or work at visionary schemes while other boys are engaged in sport. Children, as well as adults, should exhibit a modicum of learning and of virtues, and possibly a few vices.*

Emminghaus describes a form of acute paranoia with hallucinations which is said to occur in children after febrile attacks, but the cases he quotes are not very convincing; they are to be distinguished from ordinary mania by the evidence of hallucinations and by their mode of onset.

The course of paranoia is eminently chronic; for a time slight remissions may occur, enabling the person to return to some regular occupation. He may be able to keep his delusions in abeyance for a varying period of time, but they will come to the front in the end. The chronic stage is established sooner or later, in which he is entirely controlled by his delusions, and is a fit subject for asylum treatment. Very little can be attempted in this disease in the way of treatment, but if the first traces of the disorder are observed in a young boy, a special endeavor should be made to provide him with healthful surroundings, to divert his mind from his own person, and to arouse his interest in those things which are the reverse of those he is accustomed to brood over. Special teachers, and tours to foreign lands may accomplish something; but disappointment to parent, teacher, and physician is the most common result.

Moral Insanity.—We need say little of this condition, which has given rise to so much discussion. It is generally conceded that a lack of the moral sense may be the chief feature, but this moral imbecility is generally associated with a defect in the intellectual sphere; hence many of the

* In this chapter the author has drawn occasionally upon the descriptions given by him in his article on Insanity and Crime, in Hamilton's System of Legal Medicine.

subjects of moral insanity are idiotic or feeble-minded persons.

Maudsley, some years ago, went out of his way to defend the rights of moral insanity. "It may be witnessed even in young children, who, long before they have known what vice meant, have evinced an entire absence of moral feeling, with the active display of all sorts of immoral tendencies, a genuine moral imbecility or insanity." But the author is compelled to add that "associated with this defect there is frequently more or less intellectual deficiency, but not always; it sometimes happens that there is a remarkably acute intellect with no trace of moral feeling."

There are some, children and adults, whose intellectual faculties are on a far higher plane than their moral qualities, and in whom the latter cannot possibly be fostered. In a family, one of five or six children may be the only one to resist the influences of religious and secular training. Many authors are firmly convinced that the defect in morality is to be ascribed primarily to an intellectual defect; opposing views are held by some of the ablest writers, including Lombroso, Maudsley, and Hack-Tuke. Krafft-Ebing urges the justice of retaining moral insanity as a clinical form, and to this there can be no objections.

Some years ago I saw a young man at a clinic who had been arrested for assault upon his mother, whom he had failed to kill. He was entirely indifferent to the charge brought against him, and when asked whether he thought it was proper to kill one's mother, answered, "You might as well kill your mother as anyone else." The young man had received no intellectual or moral training, no religious instruction, had grown up among the most degenerate classes, and had never received the most ordinary moral teachings. Naturally the moral sense was deficient. Such a condition is embraced in Mendel's definition of moral insanity as that form which is either congenital or acquired in the earlier years of life, and is characterized by imbecility associated with a morbid tendency to immoral actions. Binswanger holds very correctly that a number of mental diseases lead to "moral idiocy."

The prognosis of the condition of moral insanity is absolutely unfavorable; nothing can be accomplished except by placing such children with moral obliquities under the stern discipline of a reformatory or an asylum.

EPILEPTIC INSANITY.*—Mental derangement associated

* *Hysterical Insanity* has been alluded to in the chapter on Hysteria (pp. 86-88). The author was at first inclined to discuss the entire disease in this division on mental

with epilepsy has been alluded to in a previous chapter. (See page 67.) It is only necessary in this connection to recall the fact that a maniacal attack may take the place of an ordinary epileptic seizure, and that such an attack is characterized by unusual suddenness and violence. Any very sudden development of mania in a child may be regarded as a symptom of possible epilepsy, even if other epileptic signs are entirely wanting. In addition to these "psychic equivalents" of an epileptic seizure, epileptic children exhibit a marked tendency to idiocy and dementia. I have known of the condition of double consciousness in an epileptic aged eighteen, but I have not seen it in younger subjects. The more pronounced symptoms of epileptic insanity are encountered in persons past the age of puberty, but Wildermuth insists that only about twenty per cent. of infantile epileptics exhibit anything like a normal mental condition.

A general psychic degeneration is a most marked feature of epileptic subjects. The disease is very common among criminals, as are other degenerative neuroses. Alcoholism in the parent is a powerful etiological factor. (Dejerine states that in 37.7 per cent. of three hundred and fifty epileptics the father was a drunkard.) Moreover, the epileptic boy is unable to attend school and falls an easy victim to bad associates.

Paretic Dementia, the serious psychosis of middle life, characterized by a progressive dementia, delusions of grandeur, and a long series of physical symptoms (inequality and immobility of the pupils, disturbances of speech, tremor of the face and hands), is so rare in childhood that we do not feel warranted in including it among the forms of insanity occurring in childhood. The youngest paretic I have seen was a man, aged nineteen years. Spitzka observed one at eighteen years, Trumbull observed the disease in a boy of twelve years, who had an attack of hemiplegia at the age of ten years. The autopsy revealed the changes of paralytic dementia. The cases thus far observed in youthful subjects did not take as rapid a course as in the adult type.

The author would make no mention of the disease, were it not for an experience he had some years since with a young man who in the course of a few weeks became exceedingly hilarious, developed the delusion that he was enormously wealthy, showed distinct tremor of speech and of hands, inequality of the pupils, and became extravagant and grandiloquent. The diagnosis of paralytic dementia was made by competent neurologists; the boy was sent to an asylum, where all his symptoms disappeared after a few weeks. He has been entirely well since.

diseases, but hesitated to do so because the marked psychic forms of hysteria are not often observed in children. The mental changes in chorea have been referred to on page 115.

MASTURBATION AND INSANITY.—There is not sufficient reason to erect a special type to be called "Masturbatic Insanity," for the habit is present in many different forms of insanity, particularly in young subjects. But the presence of this etiological or complicating condition is always noticeable, and leaves a distinct impression upon the development of the various psychoses. In many cases of acute derangement in the young, in the various forms of dementia præcox, it is often an important and grave factor. The author has had under his observation young masturbators who at the ages of ten and twelve years displayed the first effects of masturbation, and at the ages of sixteen to twenty years presented the typical symptoms of hebephrenia (mental enfeeblement, silliness, depression, general restlessness, and irritability), and gradually developed a striking dementia. There is such a marked resemblance between various forms of insanity due to masturbation that one is sorely tempted to constitute them a special group. The following account is typical of many the author could give: At the age of twelve or fourteen years, if not earlier, the boy either instinctively or through the force of bad example begins the habit. For a time it has no marked effect upon him; he continues in his studies, but does not get on quite as well as he formerly did. For this he finds all sorts of excuses. He is supposed to be, and often is, subject to severe headaches; is tired in the morning, late in rising, loses his pleasure in out-of-door sports. If detected at this stage and convinced of the viciousness of his habit he may make a determined effort to stop; sometimes he succeeds; more often he fails. From this period on, lack of concentration upon any mental effort, general irritability, disturbed sleep, loss of flesh, are the prominent symptoms. Morbid conceptions are often troublesome, and at times also lead to delusions of persecution. A young man under my care who would gaze for hours upon a clock heard it say, "De-tec-tive, De-tec-tive," and imagined himself run down by them, but was even then too stupid to give any reason for such persecution. The patient leads an aimless, silly existence; will pore for hours over books without reading them; does nothing of his own initiative, but if forced to walk

or to take exercise of some sort does everything in a mechanical fashion. If he be the son of wealthy parents, the effort is made to distract him by travel, but all is in vain; he takes no interest in anything and simply watches for the opportunity to indulge in masturbation. Complete idiocy and dementia are the ultimate result. Reference has been made to boys, but girls also fall victims to the habit, and the symptoms they present are similar to those specified before. The prognosis of all forms of insanity due to masturbation is extremely unfavorable. I have succeeded in checking the habit in relatively few cases. Many patients continue on their evil course in spite of all precautions. It is, if anything, more difficult to control the habit in girls than in boys. If the person's reason and sense of shame can be appealed to the chances of curing him of the practice are best; the previous mental calibre is an important factor; the successful cases I have seen have been in college boys and other bright lads. If the habit has been acquired by imitation it can be checked more readily than if it is the result of an innate instinct, and the sign of a degenerated mental state. Extreme watchfulness and the liberal use of the actual cautery to the spine and even to the genitals are the only possible means of effecting a cure.

The prognosis of insanity in children has been discussed with reference to the several types. It is well to emphasize the fact once more that mental derangement in childhood is recovered from more frequently than is the case with the insanity of later years.

A word in addition to what has been said on the treatment of the various types of insanity. The doctrines of heredity and of degeneration are taking a firm hold of the medical mind. Psychologically they may be true enough, but no one has yet proved that morbid hereditary influences cannot be overcome or counteracted. If a child has an unfortunate hereditary predisposition to disease, give it every possible advantage which children without any hereditary taint enjoy. The separation of children from hysterical, epileptic, or otherwise degenerate parents, is not insisted on often enough.

CHAPTER XXX.

IDIOCY AND IMBECILITY.

By idiocy we designate a permanent and complete impairment of all the intellectual faculties. Imbecility and feeble-mindedness denote lesser degrees of mental deficiency. Both these terms are used to designate mental deficiency in a brain not yet fully developed, whereas the term "dementia" is applied to those states in which all the faculties are lost after they have been normally developed. Such dementia may occur at times in relatively young persons.

Idiocy is of special interest, not only because of its frequent occurrence, but also because it demonstrates an extreme psychic condition due, in many instances, to tangible changes in the brain. In this respect it is not unlike general paresis, and the mental conditions included under these two terms are practically the only two psychic diseases whose morbid substratum is tolerably well known at the present time. In one, the highest structures of the brain have become disorganized and are undergoing dissolution after having attained normal development, while in the other defective growth or involution takes place before a full development has been reached.

In former years idiots were much neglected; they were considered an unfortunate class for whom but little could be done, and whose actual condition was scarcely worth studying; but during the past twenty-five years or more an entire change has taken place and it is fair to say that no class of patients has been more frequently the subject of study than those with defective cerebral development.

The earliest impetus was given in this direction by the French alienists, chief among them being Esquirol; a little latter the elder Seguin called attention to the possibilities of improvement in these patients and to the proper methods of training; and within the past decade or two, the interest in the morbid conditions underlying idiocy has led to numerous able contributions.

Among English authors the names of Crichton Browne, West, Maudsley, and Langdon Down, of Clouston, Ireland, and Shuttleworth, deserve special mention; in France, Bourneville has been the most successful student of idiocy; in Germany, the works of Schüle, of Krafft-Ebing, of Emminghaus, and above all, of Griesinger, have contributed most to the advance of our special knowledge of this subject, and in this country idiocy has been studied carefully by Ray, Seguin, Mills, Kerlin, Spitzka, Hurd, Brush, Peterson, Barr, and the author.

CLASSIFICATION.—The discussion of idiocy has been not unlike that of insanity. Each author has felt called upon to subdivide the subject and to establish a rational classification. The result has been that no two authors have entirely agreed. One has attempted to give a classification based upon the etiology of the condition, another upon the pathology. Down has attempted a division based upon ethnic standards, the Mongolian, the Malay, the Indian, and the Ethiopian types.

For purposes of convenience we will divide the subject as follows:

I. Hereditary Idiocy. { (a) Congenital. (b) Developmental.

II. Acquired Idiocy.. { After brain or skull injuries (including birth idiocies). After convulsions. After infectious diseases.

III. Myxœdematous Idiocy.

I. (a). *Hereditary Congenital Idiocy* includes those conditions in which the brain at birth is deficient, although this defect need not be apparent for some weeks or even months after birth. In the chapter upon Arrested Cerebral Development I have enumerated some of the anatomical conditions with which idiocy is associated, and to this part of the subject we shall recur later on. These congenital deficiencies may be the result of disease during the intrauterine period.

Congenital idiocy is a common occurrence in families with marked neurotic taint. Parents who suffer from some form of insanity, from hysteria, epilepsy, or chorea are apt to engender idiots. Among other predisposing conditions alcoholism and syphilitic infection of either parent are by far the most frequent. The importance of alcoholism in

particular cannot be overrated. The alcoholic habits of the father at the time of procreation are surely a potent factor. Syphilis of the parent must be taken into account. Premature delivery is not uncommon in syphilitic cases, and an imperfectly developed brain at the time of birth may be the cause of idiocy.

Intermarriage is supposed by many to be a direct cause of idiocy in children, and by others the influence of such marriages is absolutely denied. The truth lies midway. In families without a hereditary taint intermarriages may be quite harmless, but if there is any, and even the slightest taint of insanity, or of any other serious nervous affection, that taint becomes intensified by such an union; and since few families are entirely free from every such tendency the rule holds good that in the majority of cases intermarriages are harmful.

Traumatism during pregnancy is another factor in the development of idiocy in the child. As the influences brought to bear are prenatal in origin, these cases may be included under the heading of hereditary idiocy; and there is all the more reason to do this as such injuries to the mother are more likely to be harmful to the normal development of a child's brain in families with neurotic taint than in families whose history is entirely negative.

I. (b) *Developmental Idiocy* includes that class of cases in which the idiocy becomes apparent at certain well-marked periods of life—during dentition, for instance, and at puberty.

II. Acquired Idiocy.—This class includes a very large number of conditions, due to the most varying accidents and diseases. First and foremost, although the cause is operative at the time of birth, we must include that large number of idiocies due to traumatism during labor. The same conditions which give rise to cerebral birth palsies (prolonged labor, instrumental delivery) also give rise to birth idiocy. Other things being equal the application of instruments is not nearly as harmful as excessively prolonged labor. In the histories of children who have become idiotic, it is often stated that the child was asphyxiated at birth; that it had frequent spasms during the earlier

weeks of life, and that it exhibited marked rigidities and palsies of the extremities.

According to Mitchell, in 57 of 494 cases of idiocy labor lasted for more than thirty-six hours; 4 of them were born with unusual haste; in 22 cases forceps were applied, and 9 of these showed the impressions of the forceps on the head after birth; 4 of them were born by version; in 6 cases there was breech presentation; 11 were twins, and 9 were born prematurely; 29 were born asphyxiated and supposed to be dead; 89 were the last children of their respective mothers.

Birth idiocy is allied to another form due to injuries to the head later in life; a fall from a bench, from a chair, from a cradle, is often sufficient to cause serious disturbance, more particularly in those children who are predisposed by inheritance to mental disease. It is possible that the easier rupture of the blood-vessels in those who have inherited a syphilitic or alcoholic taint may account for the fact that while some children escape injury after severe falls others are seriously affected by relatively slight accidents.

Among the causes of acquired idiocy none is more important to my mind than convulsions. The entire development of a child may be normal until a convulsion occurs, whether as a result of some intestinal derangement or as the precursor of an acute infectious disease. From this time onward mental decadence sets in, and a child that was previously healthy and of normal mental development begins to exhibit more and more marked mental defects until it reaches the condition of complete idiocy, in which it may remain for many years. There is no difficulty in explaining this occurrence, for the tremendous venous stasis that occurs during the acme of a convulsion is sufficient to cause rupture of the pial blood-vessels, and a very considerable hemorrhage may follow; meningo-encephalitis, terminating in a general sclerosis, is the natural result and the direct cause of the mental defect. The acute infectious diseases are often responsible for the development of idiocy. In what manner this cerebral change is brought about is not easy to state, but I believe that the cases thus produced are relatively few, and that of these some are due to an in-

fectious encephalitis, and others to the convulsions accompanying these acute infectious diseases.

Idiocy is developed not infrequently after acute meningitis early in childhood. Idiocy, blindness, deaf-mutism—all these conditions separately, and sometimes conjointly, are the unfortunate results of early meningeal disease. The explanation of this condition is relatively simple, for the meningitis and the meningo-encephalitis, if they last long

FIG. 135.—Hydrocephalic Idiot. (Peterson.)

enough, may lead to a general sclerosis and atrophy of the cortical substance, which will prevent the further normal development of the brain.

The association of idiocy with hydrocephalus is common, but as this condition is generally a secondary state, both the idiocy and the hydrocephalus are the result of the primary disease. (See Fig. 135.) With the gradual increase of the hydrocephalic fluid, however, the cortical function naturally becomes impaired; there are therefore few children with unusually large heads whose mental condition is at all normal, though it is quite remarkable to what extent the hydrocephalus may increase before an absolute aboli-

tion of function takes place. This is true not only of the general mental condition, but also of the special sensory functions of the cortex.

Idiocy and epilepsy are intimately associated with one another. In all cases in which epilepsy has been developed early in life, or in which the epileptic seizures are frequent, there is a natural tendency to mental deterioration. In the adult we speak of epileptic dementia, in younger persons of epileptic idiocy. There are, no doubt, gross changes in the brains of such subjects, and in many the seeds of both the epilepsy and the idiocy may be traced to injury either at birth or during the first years of life. Thus not only the idiocy but the epilepsy, as well as the palsy, may be the result of meningeal hemorrhage occurring during labor or in very early childhood.

Symptoms.—The chief characteristic of idiocy is the lack of ordinary mental conceptions. The brain is not able to receive impressions from the outer world, and if such are received, it is not able to utilize them in anything like normal fashion, nor to form concepts or judgments. In some instances the brain represents an entire blank, in others a few impressions have been received, and these have been developed into imperfect concepts. This absolute mental deficiency is present in typical idiots, but there are varying degrees of mental deficiency or of mental development in imbeciles or feeble-minded persons.

It is scarcely necessary to cite cases in order to establish the clinical features of complete idiocy, for they are too well known even to laymen to need elucidation.

If idiocy is complete the child or the adult may not be able to recognize its own parent. He fails to recognize any object or the use of such, and of course is not able to understand or appreciate what is said to him. In extreme instances the idiot is practically nothing more than a highly organized vegetating organism, truly animal-like in all his actions without the slightest trace of human intellect. Through faulty development of speech the defect in the general mental condition of the child first becomes noticeable. If there is a lesser degree of idiocy, the child may master a few concepts, may understand a few uttered

sounds, or may be able to pronounce such simple words as mamma, papa, and the like. He may recognize the use of objects, may be capable of slight training and instruction, so that the ideas of cleanliness, and of mine and thine may be impressed upon his mind.

From these rather marked forms of idiocy and mental imbecility there is every possible gradation until we reach those types in which speech is only a little deficient, but the mental horizon is evidently narrowed down so that relatively few concepts are formed, abstract ideas are almost entirely wanting, and the imperfect mental status is determined by the lack of ordinary motives for action, by the awkwardness in intercourse with other people, and above all by the inability of the individual to cope with others in the struggle for existence. The inferiority of the individual is discovered not in the family circle in which all conditions are favorable, but when the child is thrown into competition with others at school. If the boy or young man is compelled to seek a living for himself, the feeble-minded youth is pushed to the wall under such conditions. That such persons, moreover, cannot appreciate the reason for their failure to make a success of life goes without saying. That they are rarely capable of developing the higher religious and moral ideas is equally true. The criminal classes are recruited largely from the category of feeble-minded persons in whom the ordinary ideas of right and wrong have not been engendered by early training.

The general deficiency in intellect is supposed to be offset occasionally by a peculiar development of the mind in some one direction. Stories to this effect have been common, and have been repeated by one author after the other; Griesinger's case has become rather famous of an idiot who was said to have constructed a perfect model of a man-of-war without having had any conception of geometrical designs. Many idiots are said to possess special talent in the use of cards. Drobisch described an idiotic boy, who after reading over a single page was able to repeat word for word, even if it was in Latin, which he did not at all understand. I have had under observation a young imbecile, a boy aged fourteen, who takes special delight in reading and remembering each sign that he passes on the streets, and he can without difficulty repeat fully two hundred names in the order in which they occur on the avenues. He has a true passion for signs, and as he passes along the streets his attention is riveted upon nothing else. Some of the most astound-

ing lightning calculators have been weak-minded, if not imbecile, in everything else excepting memory of figures. It is not fair, however, to turn this about and to claim that every lightning calculator must necessarily be an imbecile, for there is no doubt that some normal minds can, by special training, reach an inordinate development in some one particular direction.

As we are interested chiefly in the mental condition of children, we must devote a little more time to the early recognition of idiocy. In cases of very marked mental deficiency the disordered condition of mind can be discovered in the first months of life. In fact if parents and physicians had a proper understanding of such conditions, the mental defect could be discovered very much earlier than it generally is. It is most instructive in this connection to consider the observations of Preyer, who, in his famous treatise on the psychic development of the child, shows how early the mental processes of a healthy child can be discerned. It is a surprise to most of us to learn from him that a child in the second day of life was able to distinguish between light and darkness; that at a very early day it appreciates the sounds of words, and that in its cooing utterances it exhibits evidence of considerable cerebration. Very few people will, of course, observe children as Preyer observed his own child, but mental deficiency can, as a rule, be made out if the parent or the physician take the trouble to compare the actions of a child with the average healthy child at the same age. It is important, therefore, to note the time of life at which children may be expected to exhibit certain signs of intelligence. (See page 7.) If children have not acquired speech at the end of two and a half years, and particularly if they fail to understand what is spoken, it is fair to infer that there will be a distinct mental defect later on.

Wildermuth has taken the trouble to tabulate a number of the more important stigmata of degeneration commonly found in idiots. Eighty-two per cent. of all idiots according to these statistics present some such signs. There were abnormal conditions of the fundus in 6 of 142 cases; malformations of the external ear in 53; abnormal position of the teeth in 32; flattening of the hard palate in 11; highly arched palate in 30; prognathism in 9; excessive thickening of the skin in 9; asymmetry of the face in 25; and abnormalities of the genital organs in 8 cases. Among the functional stigmata Wildermuth refers to anomalies of the tendon reflexes in sixty per cent.; inco-ordination of the lower extremities in ten per cent.; and in six per cent.

squinting occurred. It is a curious fact that these signs of degeneration are present not only in the truly congenital forms of idiocy, but also in those which have been acquired some time after birth.

The mere appearance of the child or of the youth will often be sufficient to lead to the suspicion of idiocy without an examination of the mental condition itself. The entire absence of speech, or of defective speech in many cases, will point the same way. Caution should, however, be exercised in not mistaking deaf-mutism for idiocy, and if a child cannot speak, careful examination should be made in other ways to determine its intelligence; not rarely, however, deaf-mutism and idiocy are combined, and at times the former has been the direct or indirect cause of the latter.

FIG. 136.—Congenital Idiot; Microcephalic Skull; Extreme Contractures of Adductor Muscles of Thighs. (From a photograph kindly furnished by Dr. Peterson.)

Among idiots the defects of skull formation are of especial interest. The microcephalic skull is particularly frequent, and is either small in all its dimensions or ample in some and very deficient in others. (See Fig. 136.) The transverse and occipital diameters may be entirely normal, indeed the horizontal circumference may be up to the average, and yet if some such heads are examined it will be found that the frontal portion is unusually small, possibly receding, while the middle and occipital portions may be entirely normal. Such discrepancies and such asymmetry should

be carefully noted, for it is more important than slight deficiencies in the total measurements. Smallness of the anterior half of the skull with receding forehead proves either that there is very little room for the brain, or that the brain being small requires very little space. Asymmetry of the skull is particularly frequent, as has been shown by Fisher and Peterson, in those cases in which idiocy is associated with early infantile hemiplegia. Another point to be remembered is that irregularities in the structure of the skull are not uncommon in healthy individuals, and that as long as the cubic contents of the cavity of the skull are near the average, defects of one kind or the other appear to play a very small part. In various parts of this book I have referred to my belief that the growth of the skull is dependent largely upon the condition of the brain within, and this accounts for the fact that in idiocy acquired after convulsions the growth of the skull ceases with the disturbed condition of the brain; microcephalus is found among acquired idiots as well as among congenital idiots.

The general restlessness of idiots is characteristic. In my lecture-room I allowed these children full sway in order to demonstrate this special feature to my class. They are about the only patients who concern themselves little about the presence of the students, and who roam about the entire room in an uneasy fashion, taking hold of everything, pulling down what they can, and always fearless of the consequences. If they cannot employ themselves in any other way, they will keep up a constant motion with the fingers, twisting and braiding them, allowing the nails to scrape the skin off the fingers. They bite their nails and are apt to tear their garments. There is frequently dribbling and drooling, and if opportunity is offered, these children are particularly fond of putting their tongues to the window-panes, or any other cold object. The general awkwardness of the movements, excitability and irritability of temper, together with a peculiarity of carriage and blankness of facial expression, complete the clinical picture of idiocy and imbecility.

Imbeciles of lesser degree often escape detection; some of them are sufficiently conscious of their defects to conceal them in the presence of others.

As they grow older the deficiency in the mental and moral make-up is evidenced in the excessive development of the animal appetites. Masturbation is developed in early years, the effect of which adds to the mental deterioration. If the sexual appetite is thoroughly aroused, gratification is sought in the most outrageous fashion, and intercourse may be attempted with children or old women. Giraud referred to an idiot who attempted to rape his own sister. All the ordinary feelings of shame and modesty are wanting. In some instances the defect is a little more marked in the emotional or moral spheres than in the intellectual. Such a condition has generally been termed one of moral imbecility; but it is always associated with a decided intellectual defect.

Weak-minded and imbecile children commit crimes either from defective judgment as to the consequences or from the simple desire to gratify their passions and impulses. The desire to witness a grand spectacle or to revenge himself upon others, has led many an imbecile to set fire to houses, regardless of all consequences.

PATHOLOGY.—We have stated that idiocy is rarely a primary condition, that it is frequently associated with other conditions which point to serious brain trouble. A complete account of the pathology of idiocy would include the terminal stage of very many different brain diseases. While it is interesting to note these various terminal states it helps us but little in determining the primary lesion. Thus Wilmarth has described one hundred brains of idiots. Among these were found conditions of sclerosis with atrophy, of tuberous sclerosis, of general diffuse sclerosis. He described, furthermore, brains in which the most marked features were degenerative changes in the vessels and higher nerve-cells. He also mentioned hydrocephalus, general atrophy, and the like, so that it would be difficult from this enumeration to make any sort of inference as to the primary morbid state. The attempt should be made to distinguish between the primary and terminal states, and thus help us to push on, however little, toward a final solution of the question. I believe we shall do best if we accept the clinical division of idiocy adopted in this chapter, and endeavor to arrange the known morbid processes as far as possible under these larger clinical subdivisions.

	Primary Lesions.	Terminal States.
Hereditary idiocy.	Large defects; hemicephalus or entire absence of a considerable portion of a hemisphere.	Same; often compensatory development of hydrocephalus with large cysts.
	Partial defects (porencephaly); most frequently in motor regions. Single or double.	Same; with addition of hydrocephalus, local cysts, and general sclerosis with atrophy.
	Small brain (microcephalus); all parts equally developed, or arrest of development more marked in frontal and occipital portions than in other parts.	Small brain, often hard and sclerotic, sometimes compensatory hydrocephalus.
	Agenesis corticalis; brain apparently normal or showing only slight changes in external configuration; maldevelopment and disintegration of cells and fibres of cortex.	Same; no sign of inflammatory conditions, blood-vessels scanty; some external hydrocephalus.
	Intra-uterine, inflammatory, or vascular disturbances, such as encephalitis, thrombosis, and hemorrhage.	Meningo-encephalitis; sclerosis, localized or diffuse; general atrophy; cysts.
Acquired idiocy. (a) Birth palsies.	Meningeal hemorrhage.	Meningo-encephalitis, local cysts, sclerosis, and general atrophy.
(b) After acute diseases and convulsions.	Meningitis, encephalitis (simple or hemorrhagic); thrombosis (specific endarteritis).	Meningo-encephalitis, sclerosis, diffuse atrophy.
(c) From traumatism.	Meningeal hemorrhage; encephalitis.	Meningo-encephalitis, abscess, sclerosis, and atrophy.

That there are other conditions which occasionally lead to idiocy cannot be doubted, but they are, I am sure, not nearly as important nor as frequent as those mentioned in the above table. If hereditary syphilis is the etiological factor it is more than likely to give rise to idiocy through the medium of local meningitis, encephalitis, or of thrombosis with its subsequent changes. Mills insists on idiocy of toxic origin, and includes under this term cases due to acute poisoning, or those following acute infectious diseases, such as measles, scarlet fever, and the like.

DIAGNOSIS.—Idiocy is recognized easily enough, except in very young children; a comparison with the attainments of other children of the same age will help to establish the fact of an inferior mental development. If a child presents, in addition to the mental symptoms, any of the physical stigmata of degeneration, the probability of its remaining

an idiot is very great. Defective development of speech after a child has passed the third year is also an important aid in diagnosis, if this condition is not the result of deaf-mutism.

THE PROGNOSIS in the majority of cases of idiocy is bad. If a child has not learned to speak, and has not acquired the simplest concepts at the age of three or four years it will remain backward for all time. In the case of imbeciles of lesser degree the prospect is not quite so gloomy. In this condition more can be expected from the careful training on the part of parents and teachers.

TREATMENT.—The treatment of idiots is, as a rule, restricted to watching over their physical development and securing for them proper hygienic surroundings. In families such children are a torment to their parents and a bad example to brothers and sisters. Their separation from home is advisable, and should be urged in spite of all sentiment to the contrary. The patients enjoy the greater liberty which special institutions afford them, and the family is better off without them.

Imbeciles and weak-minded children require more careful consideration. By the most diligent training at the hands of experienced persons much can be done for them to bring their behavior and their physical condition as closely as possible to that of the average child. I am a thorough believer in pedagogic methods and of such only. If at all possible, such children should be intrusted at a very early day to a competent teacher, who will take the time to study the peculiarities of the individual, and who will fasten upon and endeavor to develop the few signs of an intellectual awakening. It is often surprising to see how much can be accomplished with these unfortunates. In some instances, however, the task is a hopeless one, and all the evidences of imbecility can rarely be eradicated. By careful calisthenic exercises, and proper gymnastic training, much can be done to avoid the outward appearance of imbecility to which parents so seriously object. If special attention is paid to the development of speech, defects in this direction can be corrected. The larger institutions, such as the Bicêtre in Paris, and the various institutions for the

feeble-minded in this country, attain a fair measure of success. But, after all, only those who possess a modicum of intellectual development can be benefited by these methods, whereas the severer grades of imbeciles and of idiots cannot be improved by them.

Cranial surgery has been looked to as a final resort. If the imbecility or the idiocy is distinctly due to a small skull, and this small skull is the result of a premature synostosis of the sutures, the surgical procedures, as practised by Lannelongue, Keen, Gerster, and others, may be allowable; but, after all that can be said on this subject has been heard, there is but little reason to expect much improvement. The slight changes that have been noticed and have been reported in children after craniectomy scarcely warrant the procedure, and, as has been intimated in a previous section, the dangers of the operation are so great that craniectomy should only be practised at the urgent request of the parents, and after every other method has been given a fair trial and has failed. In those patients in whom there is evidence of a defective cerebral development, independently of the condition of the skull, any operation is, on *a priori* grounds, entirely useless.

Medication by drugs is rarely beneficial; yet some physicians will undoubtedly be impelled by the evidence of an existing anæmia, of rickets, or of hereditary syphilis, to exhibit the drugs commonly used in these conditions.

III. MYXŒDEMATOUS IDIOCY—SPORADIC CRETINISM.

This condition, which was well described by Bourneville many years ago, deserves special mention. The mental condition is a very striking feature of these children, but it is simply a part of the general stunting of the physical and mental growth. The condition itself, although a very rare one, is of more interest at the present day in connection with the satisfactory studies that have been made regarding the cause and the treatment of myxœdema. I have associated myxœdematous idiocy with sporadic cretinism, because the two seemed to me to be very closely related, the cretin representing, however, a more intense development of the affection than is met with in those cases described as myxœdematous idiocy by Bourneville. Cases of sporadic cretinism in every way resembling the endemic cretinoid cases, so common in the mountainous districts of Europe, occur also in a few American regions, as in the mountains of Vermont and California. The cases which we

have the opportunity to see in the larger American cities are from the foreign element of the population, and though we speak of them as cases of sporadic cretinism, they occur generally in descendants of families who have lived in regions in which cretinism has been endemic.

The child, whose picture is reproduced in this chapter, is a typical case of this kind. The boy was born of healthy Prussian parents, who immigrated to this country some seven or eight years ago. He has always been healthy, but ever since he was a year and a half of age, the parents have been struck by the fact that he did not seem to continue growing as the other children in the family did. This stunted physical growth was associated with an impairment in the development of speech and of all other faculties.

FIG. 137.—Case of Myxœdema with Idiocy. Patient Twelve Years Old; Dwarfish in Stature.

The child might not unnaturally, at the age of twelve, be taken to be three, or not much more than three years old. The boy has learned to speak a few words, is able to call his parents and some near relatives. His affections are well developed, as was evidenced when he was separated from his father and placed in the hospital, and he shows considerable aptitude in playing with other children, but not quite as much as a healthy child of three years of age would. As for his mental condition, it is not equal to that of a child of three years; beyond the expression of his simple wants, he exhibits remarkably few signs of mentality.

The disease is characterized by a peculiar glossy appearance of the skin, prominent lips, receding forehead, and a peculiar, stubbed nose. Compared with the rest of the body the stomach is inordinately large; the tongue is relatively thick, and very often protrudes from the mouth. All children with myxœdematous disease so closely resemble each other that they might be supposed to belong to one and the same family. The examination of the neck fails to reveal any trace of the thyroid gland.

ETIOLOGY.—The etiology of myxœdematous idiocy, as well as of spora-

dic cretinism, is practically unknown, except that it is very apt to occur in mountainous regions, and particularly in regions in which the water contains a great deal of salts of lime and magnesia. But this is evidently not the only explanation, for the children of persons who have emigrated from such districts have often been affected with the disease although subject to entirely different atmospheric and tellurian conditions. The frequent intermarriages between people in isolated mountainous districts also has its bearing upon the hereditary transmission of these diseases.*

DIAGNOSIS.—Myxœdematous idiocy cannot very well be mistaken for any other condition. It is only in the earlier years of life, when the stunted growth and the peculiar expression of the child are not yet fully marked, that the true cause of the idiocy may not be suspected. But the arrest in the general growth and the peculiar appearance of the skin will give the clew to the true state of affairs.†

PROGNOSIS.—The prognosis of this condition was extremely grave until very recently, and therapeutic experiments are now proceeding which will prove whether this one class of idiots may possibly be benefited by treatment. Until a decade or more ago there was as little hope of improving myxœdematous patients as of helping other congenital idiots, and a removal from the mountainous region in which cretinism was endemic, or an entire change of surroundings, was supposed to be the only possible way of procuring some relief, though actual cure of any such condition had not, to my knowledge, been recorded by anyone.

As will be seen in the discussion of the treatment of this condition there is some hope of improving these children by the use of recent methods.

TREATMENT.—All the older suggestions with regard to treatment can be discarded. Feeding the patient with the thyroid gland promises some relief. The author has been personally interested in the treatment of several patients with myxœdematous idiocy. In one of them a very decided change was effected, the mental and physical symptoms being greatly benefited by the use of thyroid extract. In two other cases the treatment was entirely unsuccessful, and in a fourth patient, a cretin, aged thirty-four years, a considerable change in the physical condition was effected, but the patient became so emaciated that the drug had to be discontinued.‡ The pulverized gland is now for sale in the market and can be obtained easily in any quantity. With children the treatment should be begun with doses of one grain of the thyroid gland, to be given three times a day, and increased from one

* For the pathology of this condition, see page 199.

† These conditions bear a superficial resemblance to infantilism—a condition characterized by stunted growth, a persistence of child-like physique, non-development of the genital organs, and of those physical and mental traits which go with normal sexual growth (Bourneville, Brissaud, Meige, Lorain, and other French and Italian writers). There is some resemblance also to achondroplasia—a stunted growth of the extremities with normal growth of the trunk (Marie). Skull relatively large. The epiphyses are deformed and thickened. Intelligence, as a rule, remains normal.

‡ All these cases were treated more than ten years ago. With the improved method of administering the "gland," better results have been obtained.

up to five grains. While the drug is being given the child should be under proper medical observation, and the effect of the administration of the gland upon the heart and other organs should be carefully noted. This treatment should be begun in the earliest stages of myxœdematous idiocy, with the view to prevent, if possible, the full development of the disease.

APPENDIX.

Localization of the Functions of the Segments of the Spinal Cord.

For reference, the following tables, as revised by Starr, are appended.

Segment.	Muscles.	Reflex.	Sensation.
II. and III. C.	Sterno-mastoid. Trapezius. Scaleni and neck. Diaphragm.	Hypochondrium (?). Sudden inspiration produced by sudden pressure beneath the lower border of ribs.	Back of head to vertex. Neck.
IV. C.	Diaphragm. Deltoid. Biceps. Coraco-brachialis. Supinator longus. Rhomboid. Supra- and infra-spinatus.	Pupil. 4th to 7th cervical. Dilatation of the pupil produced by irritation of neck.	Neck. Upper shoulder. Outer arm.
V. C.	Deltoid. Biceps. Coraco-brachialis. Brachialis anticus. Supinator longus. Supinator brevis. Rhomboid. Teres minor. Pectoralis (clavicular part). Serratus magnus.	Scapular. 5th crevical to 1st dorsal. Irritation of skin over the scapula produces contraction of the scapular muscles. Supinator longus. Tapping its tendon in wrist produces flexion of forearm.	Back of shoulder and arm. Outer side of arm and forearm, front and back.
VI. C.	Brachialus anticus. Serratus magnus. Triceps. Extensors of wrist and fingers. Pronators.	Triceps. 5th to 6th cervical. Tapping elbow tendon-produces extension of forearm. Posterior wrist. 6th to 8th cervical. Tapping tendons causes extension of hand.	Outer side of forearm, front and back. Outer half of hand.
VII. C.	Triceps (long head). Extensors of wrist and fingers. Pronators of wrist. Flexors of wrist. Subscapular. Pectoralis (costal part). Latissimus dorsi. Teres major.	Anterior wrist. 7th to 8th cervical. Tapping anterior tendons causes flexion of wrist. Palmar. 7th cervical to 1st dorsal. Stroking palm causes closure of fingers.	Inner side and back of arm and forearm. Outer half of the hand.
VIII. C.	Flexors of wrist and fingers. Intrinsic muscles of hand.		Forearm and hand, inner half.

LOCALIZATION OF THE FUNCTIONS OF THE SEGMENTS OF THE SPINAL CORD.—*Continued.*

Segment.	Muscles.	Reflex.	Sensation.
I. D.	Extensors of thumb. Intrinsic hand muscles. Thenar and hypothenar eminences.		Forearm, inner half. Ulnar distribution to hand.
II. to XII. D.	Muscles of back and abdomen. Erectores spinæ.	Epigastric. 4th to 7th dorsal. Tickling mammary region causes retraction of the epigastrium. Abdominal. 7th to 11th dorsal. Stroking side of abdomen causes retraction of belly.	Skin of chest and abdomen, in horizontal bands running around the body.
I. L.	Ilio-psoas. Sartorius. Muscles of abdomen.	Cremasteric. 1st to 3d lumbar. Stroking inner thigh causes retraction of scrotum.	Skin over groin and front of scrotum. Over back above buttocks.
II. L.	Ilio-psoas. Sartorius. Quadriceps femoris. Flexors of knee.	Patella tendon. Striking tendon causes extension of leg.	Front of thigh.
III. L.	Quadriceps femoris. Flexors of knee. Inner rotators of thigh. Abductors of thigh.		Front and outer side of thigh.
IV. L.	Abductors of thigh. Adductors of thigh. Tibialis anticus.	Gluteal. 4th to 5th lumbar. Stroking buttock causes dimpling in fold of buttock.	Outer side of leg. Inner side of thigh and leg to ankle. Inner side of foot.
V. L.	Outward rotators of thigh. Flexors of ankle. Extensors of toes.		Back of thigh, back and outer part of leg and of foot.
I. to II. S.	Flexors of ankle. Long flexor of toes. Peronei. Intrinsic muscles of foot.	Plantar. Tickling sole of foot causes flexion of toes and retraction of leg.	Back of thigh and outer side of foot. Buttocks.
III. to V. S.	Perineal muscles.	Foot reflex. Achilles tendon. Over-extension of foot causes rapid flexion; ankle-clonus. Bladder and rectal centres.	Skin over sacrum, anus, perineum, genitals, and lower part of buttocks.

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